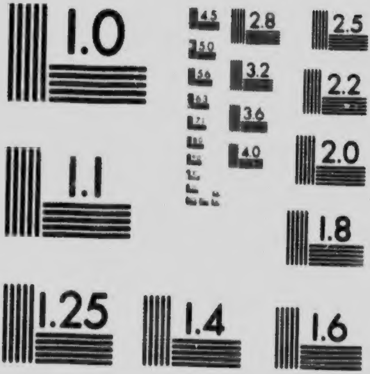

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OTOSCLEROSIS

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(IDIOPATHIC DEGENERATIVE DEAFNESS)

BY

lexander

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INTRODUCTION

If a number of experienced otologists were asked to express the value they attach to the various signs and symptoms by means of which the diagnosis of otosclerosis is made, there would probably be found to be wide divergences of opinion. And yet, if these same observers were asked to make a clinical diagnosis of a large number of cases of deafness, I venture to think that there would be remarkable agreement in respect to the cases which they would class under the term otosclerosis. I would even go further than this, and am strongly of the opinion that were these cases made the subject of pathological examination, it would be found that the diagnosis in the great majority of cases would be correct.

The reason for this is probably the fact that, consciously or unconsciously, the experienced otologist will not allow his judgment to be swayed by the presence or absence of any one special symptom, because he knows that its significance may be ambiguous. His diagnosis rests on the clinical picture which the case presents from its mode and age of onset, its history, the family history, the appearance of the drumhead, the absence of signs or symptoms indicating involvement of the functions of equilibration, the tests for hearing, the diminished sensitiveness of the skin of the external meatus and drumhead, the effects of inflation, etc. But

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on no single one of these signs and symptoms would the experienced otologist place much reliance. The only condition that leads to a common error in diagnosis is the evidence of present and, still more often, of past suppurative middle ear disease. It is a curious fact that it is very rare to hear of a diagnosis of otosclerosis in a case in which there is a perforation of the membrane and other evidence of suppurative middle ear disease. Apparently the mind of the aurist is satisfied that the visible changes are sufficient to account for the symptoms. This, at any rate, I may frankly state is the error I most frequently made until I made a more careful study of the disease. Then I found that the combination of the two conditions was by no means uncommon.

The difficulties associated with the otosclerosis problem, therefore, are not so much those of symptomatology and diagnosis, but rather those of its pathogenesis, its pathology and biological significance, and its treatment. It is for this reason that in this work I have directed attention rather to the latter aspects of the question than to those of diagnosis and semeiology. The conclusions I have drawn from the consideration of the clinical features of the disease, the pathological anatomy, and the hereditary element present in many cases inevitably lead to the subject of the nomenclature of the disease. The word otosclerosis is too obviously a misnomer. For reasons given in the text I have ventured to suggest the term Idiopathic Degenerative Deafness.

All the photographs and photomicrographs have been taken by myself, and I have not allowed any " touching up " in the prints. When flaws have been present in the sections, I have allowed them to appear in the photographs. This no doubt diminishes the artistic appearance of the illustrations, but it is most important from the reader's point of view that he should not be deliberately misled in his conception of the pathological facts.

The pathological research was carried out by myself in the pathological laboratory of the Victoria Infirmary, Glasgow.

ALBERT A. GRAY.

May 14th, 1917.

CONTENTS

PAGE
INTRODUCTION v

CHAPTER I

AGE AND SEX 1

CHAPTER II

HEREDITY AS A FACTOR IN THE OCCURRENCE OF OTOSCLEROSIS 5

CHAPTER III

BIOLOGICAL 49

CHAPTER IV

PATHOLOGICAL ANATOMY 66

CHAPTER V

GENERAL CONSIDERATIONS ON THE PATHOLOGICAL CHANGES 142

CHAPTER VI

NOTES ON THE TREATMENT AND PROGNOSIS OF OTOSCLEROSIS 163

BIBLIOGRAPHY 190

LIST OF ILLUSTRATIONS

FACING PAGE

CASE I

RIGHT EAR:

FIG. 1. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE 82
FIG. 2. SECTION THROUGH THE FOCUS OF DISEASED BONE 84
FIG. 3. SECTION THROUGH THE LINE OF DEMARCATION AND ADJACENT REGIONS . . . 86

LEFT EAR:

FIG. 4. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE 88
FIG. 5. SECTION THROUGH THE FOCUS OF DISEASED BONE 90

CASE II

RIGHT EAR:

FIG. 6. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE 98
FIG. 7. SECTION THROUGH THE FOCUS OF DISEASED BONE 100

LEFT EAR:

FIG. 8. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE . 102

FACING PAGE

CASE III

RIGHT EAR:

FIG. 9. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE . 110

FIG. 10. SECTION THROUGH THE FOCUS OF DISEASED BONE IN FRONT OF THE OVAL WINDOW 112

FIG. 11. HORIZONTAL SECTION THROUGH THE STAPEDIO-VESTIBULAR ARTICULATION . . 114

LEFT EAR:

FIG. 12. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE 116

FIG. 13. SECTION THROUGH THE FOCUS OF DISEASED BONE 118

FIG. 14. SECTION THROUGH THE STAPEDIO-VESTIBULAR ARTICULATION 120

CASE IV

LEFT EAR:

FIG. 15. STEREOSCOPIC VIEW OF THE INNER WALL OF THE TYMPANUM 130

FIG. 16. HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE 132

FIG. 17. SECTION THROUGH THE ANTERIOR REGION OF THE STAPEDIO-VESTIBULAR ARTICULATION 134

FIG. 18. SECTION THROUGH THE POSTERIOR MARGIN OF THE OVAL WINDOW AND ADJACENT STRUCTURES 136

FIG. 19. SECTION OF THE RAREFIED BONE IN THE REGION BEHIND THE OVAL WINDOW . . 138

FIG. 20. SECTION OF A PORTION OF THE RAREFIED BONE, SHOWING OSTEOCLAST . . . 140

OTOSCLEROSIS

CHAPTER I

AGE AND SEX

It is universally accepted by aurists that otosclerosis first manifests itself most commonly between the twentieth and the fortieth year. My own investigations fully corroborate this, and indeed I have found that the first incidence of the disease is most common between the ages of twenty and thirty. My statistics reveal a rather larger percentage of cases arising between the ages of ten and twenty than do those of some others, *e.g.* Hartmann,[1] and although the disease is very rare before the age of ten considering the large proportion of the population alive during that first decade, still cases do occasionally occur then, as may be seen from the accompanying table. In this connection it is interesting to note that judging from my own experience, cases of otosclerosis arising in childhood are generally found to occur where there is the chance combination of two factors—a marked inherited tendency to otosclerosis associated with suppurative middle-ear disease.

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[1] Hartmann, *Ueb. d. Knöchern Fixation d. Steigbügel*, s. 44 (J. F. Bergman, 1898).

TABLE I

SHOWING INCIDENCE OF OTOSCLEROSIS IN REGARD TO AGE AND SEX

Age	0–10.	10–20.	20–30.	30–40.	40–50.	50–60.	Over 60.
Male	2	6	9	3	2	4	0
Female	4	18	30	12	4	1	0
Total	6	24	39	15	6	5	0

In studying the accompanying table, the reader must remember the very important fact that the total population in any given decade is considerably less than that of the preceding decade, and greater than that of the succeeding one.

Thus the six cases of otosclerosis originating in the decade 0–10 is a very small percentage of the total number living in that decade, as compared with the six cases originating in the decade 40–50.

Similarly, the ratio 24 : 39, in the second and third decades respectively, does not give a correct relationship to the actual number of the population living. When corrected it would be found that the interval in the ratio would be considerably greater even than it appears.

On the other hand, the drop in the frequency of the incidence of the disease between the decades 30–40 and 40–50 is not really so great as the ratio 15 : 6 would seem to indicate.

For a totally different reason there is probably an error in the ratio 6 : 5 between the decades 40–50 and 50–60 respectively. We would expect the interval in the ratio to be more than that, and it is probable that an error is present in this case, because the numbers upon which to attempt to base a numerical relationship are too small.

Sex.—As regards sex it will be seen that out of the ninety-five cases, twenty-six occurred in men and sixty-nine in women. This indicates a considerably higher incidence in women than most previous investigators have found. It is possible that an error may be present in my own figures owing to the number available, ninety-five, being too small for statistical purposes. On the other hand, the statistics of some of the earlier investigators, such as Bezold and Hartmann, may perhaps have been vitiated by an over-stringency in the diagnosis of the disease, which more recent observation has shown to be unjustifiable. In other words, the earlier investigators were making a diagnosis, not of otosclerosis, but of fixation of the stapes

As regards the relative sex incidence in the different decades, a very interesting fact is revealed in Table I. Taking the decades in their age order and reducing the figures in such a way that the male incidence is represented by unity throughout, while the numbers represented by the female incidence is variable, we get the following ratios in the different decades respectively :

1 : 2, 1 : 3, 1 : 3·2, 1 : 4, 1 : 2, 1 : 0·25.

From this we see that in all the four earlier decades of life not only is the frequency of otosclerosis greater in women than in men, but that the ratio of its frequency in women undergoes an increase. In the fifth decade, although the actual frequency of the disease is greater in women, yet the ratio undergoes a sudden drop. Finally, in the sixth decade, the ratio between the two sexes actually becomes reversed, and men begin to suffer more frequently than women. It must, however, be pointed out that in regard to the fifth and sixth decades, the numbers available in the table are very small, and too much importance must not be attached to them. They indicate, however, a line for future research.

In general these figures agree with those of other investigators, and they show that between the ages of ten and twenty changes take place in the body which increasingly determine the occurrence of otosclerosis. These changes become still more pronounced between the ages of 20 and 30, after which they appear to diminish. It is very improbable that only one specific change is the immediate cause of the incidence of otosclerosis. The occurrence of puberty, the cessation of growth, the completion of the processes of ossification, and of the development of the nervous system may all play their part in various degrees in different cases as causative factors.

It must again be pointed out that the numbers given above are not very large, and therefore some allowance for errors must be made. The subject is an interesting one, and with larger numbers could be made a special and valuable line of investigation, provided reasonable care was exercised in the selection of cases so that only those of true otosclerosis were included.

CHAPTER II

HEREDITY AS A FACTOR IN THE OCCURRENCE OF OTOSCLEROSIS

THE influence of heredity in the causation of deafness has now been recognised among aurists for very many years, and it is highly probable that long before otology as a branch of medical science had come into existence, some knowledge of such a relationship was common even among the laity.

When the study of ear affections was more definitely undertaken, the influence of heredity was recognised. Further, the fact was supposed to be clearly demonstrated that the relationship existed only between heredity and that form of deafness which is not characterised by discharge in the middle ear or by the pathological changes which resulted from such conditions. In other words, it was only among cases of the affection known as " dry catarrh " that heredity was admitted to be a factor in the condition. Such a belief is widely held to-day, but, as will be shown later, there is every reason to think that this rigid view must be abandoned.

With the gradually increasing knowledge of the anatomical changes which are associated with deafness it became clear that the term " dry catarrh " was unfortunate, and included several different pathological conditions. A complete knowledge of the changes which occur in these cases is not yet in sight, but sufficient advance has been made to show that, in a considerable proportion of them at least, the changes take place not only in the middle ear (and in many cases not in that cavity at all), but more particularly in the bony capsule of the labyrinth. Sometimes associated with these are found changes in the membranous structures of the labyrinth and in the auditory nerve. These cases now fall under the category of otosclerosis.

It is curious to observe that while considerable interest was taken in the pathological changes, the relationship which was known to exist between heredity and otosclerosis excited little further remark. With the exceptions to be noted later, the only works of reference I can find on this subject relate to the percentages of cases which are inherited compared with those which are not. Now, it will be shown that any such attempts to draw a distinct line between cases of otosclerosis which are due to inherited tendency and those which are not, are doomed to failure. I, myself, have recently tried to estimate these percentages in my own cases, but when the family and personal histories came to be investigated carefully, it became quite evident that no sharp line could be drawn between the two. In otosclerosis we can only speak of inheritance being present in greater or less degree. This important aspect of the question will be referred to again after the cases have been described in detail.

Reference has just been made to the comparative absence of investigation in regard to the relationship between heredity and otosclerosis. Exceptions to this statement must be admitted in the case of certain otologists in recent times, and their work will be quoted in the following pages. Speaking generally, however, there has been little accurate investigation, and this is probably due to the following causes. First, the unfortunate dominance that the study of bacteriology gained over pathology in its larger meaning. The micro-organism and its products were studied with the utmost zeal, while, with the exception of the leucocytes, the response which the living elements of the body made to the bacteria and their poisons as well as to other injurious agents was for the most part neglected. The inevitable result followed. The clinical facts of the disease observed by the physician and surgeon did not fall into the cut-and-dried categories which the bacteriologist had provided for them. Hence it came about that, in respect to diseases in general and among them otosclerosis, the study of heredity as well as several other important subjects was somewhat neglected. Another factor which deflected the attention of otologists to a large extent from the consideration of heredity and other matters in relation to the ear, was the phenomenal outburst of surgical activity. This activity was, of course, highly beneficial on account of the valuable practical advances which resulted from it. At the same time it drew away much attention from the study of non-suppurative diseases of the ear, and it has left arrears which the present and future generations of otologists will have to fill up.

But there is another factor in special reference to the effect of hereditary influence in otosc[illegible] which no doubt has deterred many from making [illegible] matter the subject of investigation. This is the difficulty of obtaining data. Those who have attempted to investigate the subject will understand well what is meant. Those who have not made any such attempt might be well advised to try to ascertain the family history of deafness in some of their patients, and they will be surprised to find how little the human subject knows of the troubles which afflicted his ancestors or still afflict his living relatives. There is usually no difficulty in finding out whether the brothers, sisters, or parents of a given patient suffer from deafness, but as regards even first cousins very often no information can be gained. Concerning uncles and aunts information can sometimes be obtained in respect to the presence or absence of deafness, and not infrequently the same is true of grandparents. Only rarely is information forthcoming concerning grand-uncles or grand-aunts. These remarks apply even to the case of patients who are anxious to give real information concerning the condition of the hearing of their relatives and ancestors. But there still remain other cases in which the difficulty is increased by the pious self-deception of the patients themselves. It appears to be an article of faith with some individuals that no qualities are inherited in their own particular family except those which are desirable. Should any reader therefore desire to make an investigation such as the present, he must never accept the patient's statement that there is no deafness in the family. The only way to get even approximately accurate data is to obtain the family tree, so far as the patient can supply it, and then make inquiries concerning each member separately. Educated people, particularly scientific and medical men, usually look upon these matters in a rational way, and their statements are consequently more reliable than those of less educated individuals.

Among those who in recent times have interested themselves in the relationship between heredity and otosclerosis, Körner and Hammerschlag have taken especial trouble in tracing out the family histories of several of their patients, and their work will be referred to later in greater detail.

The method employed in this book is, first of all to give, so far as is known, the family tree of the patient or patients, those who are or were deaf being printed in black squares, and those not so affected in white ones. The conventional symbols ♂ and ♀ are used to denote male and female respectively, the different generations are marked A, B, C, etc., and the individuals in each are numbered in each family. When possible, the details of each case are given below the corresponding tree. This last aspect of the subject has hitherto been almost entirely neglected. It was, indeed, only undertaken by myself for the purpose of making the investigation as complete as possible, and was not expected to give much tangible result. As the subject has evolved itself, however, I have found that from this detailed history one of the most interesting results of the investigation has accrued.

It is, therefore, for this reason that I have not contented myself with merely giving the family trees, but have, where possible, added notes on the individual cases in regard to their history and the condition of the hearing at the time of examination. In this way the reader can judge for himself the extent to which, in each case, hereditary influence has been at work on the one hand, and extraneous factors on the other.

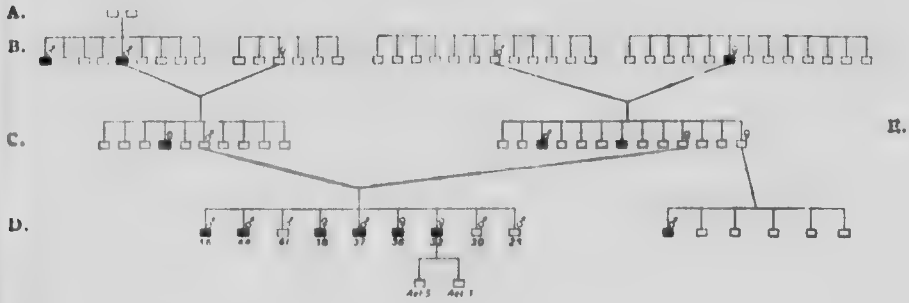
A word must be said as to the standards used in testing the hearing, in order that the reader may judge of the degree and nature of the deafness. As regards hearing distance, three standards were employed—the watch, the whispered voice, and the conversational voice. The same watch was employed throughout all the examinations, and the standard is therefore constant. It is heard by a normal ear at a distance of from 2 to 2½ yards. The whispered voice is heard by a normal ear at a distance of between 6 and 7 yards. The conversational voice, as heard by a normal ear, cannot be correlated with the same as heard by a defective ear, because it is heard normally at a distance far greater than is possible to obtain in the rooms in which the examinations were carried out. In the following pages, therefore, it is only of use in comparing one defective ear with another, and when it is heard at a distance of over 8 yards the sign + is prefixed in order to indicate that the distance is more than that, but how much more is not known.

The Rinne and Weber bone-conduction tests were all carried out by the same fork A^1 (220 v.d. per sec.), and the result of the former test in a normal hearing ear is between + 12 secs. and + 15 secs. The fork employed throughout for Schwabach's test was of the same pitch (220 v.d.), and, when placed on the mastoid process of the normal adult ear, is heard for 17 secs. For the testing of the lower notes a series of forks made by König was employed, and the French notation is therefore given.

The Edelmann-Galton whistle was used for the purpose of testing the high notes. In this connection it must be remembered that the vibration-frequencies, which are given in the table accompanying this instrument, have been called in question by some very good authorities. In order, therefore, to eliminate the possibility of errors arising from this source, I have not put down the notes according to their supposed frequencies, but have indicated the loss of hearing in terms of the numbers on the instrument which give the length of the hollow pipe and the distance of the mouth-piece from the latter. Thus, when it is stated in any given case that the notes are lost above Mdh. (Mundhöhle) 6·0, Pfl. (Pfeifenlänge) 3·4, it means that, when the lower and upper screws on the instrument are placed at these numbers respectively, the note produced is the highest which the patient can hear. I have not employed Gellé's test in this investigation as I have never been able to satisfy myself that it is reliable.

The first family tree is the most complete and instructive that I have been able to procure. (See Table II.)

TABLE II



Viewing the tree as a whole, it will be seen that both on the paternal and maternal sides fecundity is a prominent feature, though probably this is not to be associated with the otosclerosis. In the next place, on the paternal and maternal sides of the present generation the families were only very slightly burdened by deafness. It must be mentioned that no blood relationship existed between the father's and the mother's families. Another very important fact to be noted is that neither the father nor the mother of the present generation were in themselves deaf, and yet, of the nine children of this marriage, six are already the victims of otosclerosis, and of the three who are not deaf, one has not yet reached the age at which her brothers and sisters tended to become affected, and another is only just at that age, viz. 28 to 30 years. The only one with good hearing who has definitely passed that critical age is at present 41 years old.

D 1. The patient is a male, æt. 46. He was examined by me and the results of examination are tabulated below :

Right Ear.		Left Ear.
− 24.	Rinne.	− 15.
+ 10.	Schwabach.	+ 7.
Not heard at all.	Watch.	Not heard at all.
9 ins.	Whisper.	Not heard.
1½ yards.	Conv. voice.	2 ft.
Mdh. 8·0, Pfl. 10·3.	Galton's whistle.	Mdh. 6·0, Pfl. 9·7
Lost below La $_1\sharp$.	Low notes.	Lost below Re$_3$.

On inflation, air passes up both Eustachian tubes freely, but produces no improvement.

The right tympanic membrane shows the presence of an old dry perforation. The left tympanic membrane is normal. No symptoms or signs of disturbance in the semi-circular canals or vestibule have ever occurred, and the caloric tests for nystagmus show that the labyrinthine reflexes are normal in both ears. Tinnitus is constant and very distressing, and is worse in the right, that is, the better hearing ear. Paracusis is pronounced. The patient is a business man, and is not subjected to any loud noises.

The first trouble with his ears began at the age of 11, when he had very severe pain followed by discharge which lasted at least some weeks. He cannot remember whether this condition occurred in both ears, but deafness was present in both, and has remained ever since. During the last ten years or so the deafness has gradually become more pronounced. At present there is no catarrhal trouble in the upper air passages, but it is quite probable that such was in existence in childhood.

D 2. Male, æt. 44. No opportunity has occurred of examining this patient. From information received from other members of the family, it is evident that he became slightly deaf about the age of 30. The deafness has not increased very much.

D 3. Æt. 41. This subject, a male, has had no trouble with his ears at all.

D 4. Female, æt. 38. The results of the examination are tabulated below:

Right Ear.		Left Ear.
− 20.	Rinne.	− 18.
+ 10.	Schwabach.	+ 8.
3 ins.	Watch.	Contact only.
5 ins.	Whisper.	Not heard.
1 yard.	Conv. voice.	3 ins.
Lost above Mdh. 6·0, Pfl. 2·3.	Galton's whistle.	Lost above Mdh. 6, Pfl. 2·9.
Lost below re$_1$.	Low notes.	Lost below ut$_5$.

No appreciable departure from the normal was discernible in either tympanic membrane, and there was no rosy tint over the region of the promontory. Air passed readily up the Eustachian tubes, and produced a slight but very temporary improvement on the right side only. Tinnitus is present and is moderately severe; paracusis is also noticeable. No symptom of disturbance in the canals or vestibule has ever been present, and the caloric tests for nystagmus give a normal response.

As regards the history of the case it is reported that the dulness of hearing began in the right ear at the age of 16, and in the left ear at the age of 23. There is no history of suppuration or of earache, but the remains of adenoids are present to a slight extent. There is no history of nasal or naso-pharyngeal trouble. The tinnitus made its appearance after the deafness, but the length of the interval between the onset of the two symptoms cannot be ascertained.

D 5. Male, æt. 37. No opportunity has occurred of examining this subject, but from the other members of the family it is learnt that he suffers from dulness of hearing and tinnitus, and that these symptoms made their appearance about the age of 28 or 29. There is no history of otorrhœa or earache.

D 6. Female, æt. 36. The results of the examination are tabulated below :

Right Ear.		Left Ear.
− 5.	Rinne.	− 3.
+ 7.	Schwabach.	+6.
3 ins.	Watch.	7 ins.
1 yard.	Whisper.	2 yards.
8 yards +.	Conv. voice.	8 yards +.
Lost above Mdh. 6·0, Pfl. 2·4.	Galton's whistle.	Lost above Mdh. 6·0, Pfl. 2·3.
Lost below ut_1.	Low notes.	Lost below si_1.

There was a considerable amount of wax present in both ears, and when this was removed it was found that the tympanic membranes were normal. No rosy tint was present over the region of the promontory. On inflation, air passed freely up both Eustachian tubes and produced a very slight and transient improvement in the right ear, but had no such effect on the left. At no time has there been any symptom of disturbance in the functions of the canals or the vestibule, and the response to the caloric tests for nystagmus is normal on both sides. Tinnitus of a buzzing character is constantly present in the right ear, but it is only rarely noticed in the left. Paracusis is clearly observed by the patient, and she states that this symptom first made its appearance a year ago.

The dulness of hearing in the right ear began at the age of 29, and in the left ear at the age of 32. There is no history of nasal or naso-pharyngeal trouble, nor of earache or otorrhœa. On examination, the upper air passages appear to be quite healthy.

D 7. Female, æt. 32, but at date of examination, Nov. 1902, was æt. 24. Since that date no opportunity has occurred of re-examining the patient. The results of the examination are tabulated below:

Right Ear.		Left Ear.
− 5.	Rinne.	− 4.
?	Schwabach.	?
1 ft.	Watch.	1 ft.
2 yards.	Whisper.	2½ yards.
6 yards.	Conv. voice.	6 yards +.
All heard.	Galton's whistle.	All heard.
A few lost.	Low notes.	A few lost.

On examination there was, perhaps, a slight indrawing of the membrane, but this cannot be definitely stated as amounting to a pathological change. No rosy tint was observed over the promontory, and the membrane was normally movable. On inflation, air passed readily up both Eustachian tubes and was followed by a very slight degree of improvement in both ears; this improvement, however, passed rapidly away. There has never been any history of giddiness or symptoms pointing to disordered function of the semi-circular canals or the vestibule. Tinnitus is present constantly in the right ear and occasionally in the left. It made its first appearance some months after the deafness. There has never been any noticeable nasal or naso-pharyngeal trouble, and the general health, like that of the other members of the family, has been good. Shortly before the onset of the [illegible]ess, however, the patient was treated for chlo[illegible], between t[illegible] and the otosclerosis there may be some relationship of cause and effect. There is no history of earache or otorrhœa.

It is interesting to note that three years after the examination reported above this patient married, and has had two children. Her sisters are of opinion that her hearing appeared to be somewhat worse after the birth of each child. No defect of hearing has been noticed in either of these two children, but, of course, they are as yet too young for the occurrence of otosclerosis.

D 8. Male, æt. 30. Hearing examined Nov. 1910, and found to be perfectly normal in every respect.

D 9. Female, æt. 25. Hearing examined Nov. 1910, and found to be normal in every respect.

Taking the previous generation, C, in the same family tree on the paternal side, it will be seen that deafness is found, but that the family is not at all heavily burdened, only one out of a family of ten being affected. The subject, C 4, is dead, and the only information that can be definitely ascertained is that she became deaf in middle life. It is important also to note that the father, C 6, of the present generation was not deaf when he died at the age of about 60.

Going back still another generation, B, on the paternal side deafness is found in the family of the grandfather. There is a granduncle, B 1, deaf, but there is evidence that this appeared fairly late in life; there was no noticeable deafness before the age of 60. The paternal grandfather himself, B 5, became dull of hearing, but this also occurred comparatively late in life. There was no deafness in the family of the paternal grandmother, out of a membership of six.

In the family of the great-grandparents on the paternal side, A, deafness so far as can be ascertained was absent, but obviously the possibility of obtaining reliable evidence is remote.

On the maternal side, E, of the present generation, it is found that deafness was present in two members, E 3 and E 7, out of a large family of thirteen; but it is important to notice that the mother of the present generation was not deaf.

Going back another generation on the maternal side, F, it will be seen that in one of the families there is no deafness out of a family of twelve. In the other family of the same generation on the mother's side, out of a family of thirteen, one is deaf, and that one is the grandmother of the present generation. In this case the deafness did not occur till the age of 50.

Finally, in respect to the cousins of the present generation, the facts are these: on the father's side there are over forty first and second cousins, and of these none is deaf. On the mother's side there are over thirty first and second cousins, and of these only one is deaf. He is a young man, aged about 30.

Summarising the facts obtained from a study of the genealogical tree, it is found that, out of a family of nine, six actually suffer from otosclerosis, and it is quite possible that all may ultimately become affected. Of the three who have so far escaped one has reached the age of 41, so he is distinctly above the age which is critical for this particular family. Still, he has not by any means passed the age at which otosclerosis occasionally becomes manifest. Of the remaining two who are unaffected, one is just passing the critical age and the other is entering upon it.

In spite of this excessive preponderance of otosclerosis in the children, it is clear that in the families of each parent taken separately the burden of deafness is only slight. The evidence of this is from two sources—first by direct inspection of the tree, and second, by the fact that out of more than forty cousins (first and second) on the father's side none is deaf, and out of more than thirty cousins on the mother's side only one is deaf.

While it is obvious that in this family there is a strong predisposition to otosclerosis, it is interesting to observe that no certain deduction can be made as to the side, paternal or maternal, from which the tendency is derived. Both sides are only very slightly burdened with deafness, and it is very doubtful, from what is known of the incidence of otosclerosis in other cases which are markedly hereditary, whether such persistent incidence as is found in the present family could be accounted for by inheritance from one unaffected parent only. In this case, I think the strong probability is that the tendency is derived from both paternal and maternal sources. This tree should be compared with that of Hammerschlag (Table IX).

Another feature which is noticeable in this family history and will be referred to later in greater detail, is the tendency for the affection to manifest itself at an earlier period of life in successive generations. Probably associated with this observation is another interesting fact to be noted in this family tree, as in others. Provided adventitious factors be excluded, there appears to be a tendency for the disease to become manifest at the same age in members of the same family. Thus, in the family tree under discussion it will be observed that out of the family of the present generation there are six deaf. Of these, three (D 2, D 5, D 6) became deaf, without any known exciting cause, at the age of 29 or 30. In another member, D 7, the disease made its appearance at the age of 24, but it is to be noted that the patient was suffering from chlorosis shortly before the dulness of hearing and tinnitus first appeared. In D 4 the deafness began at the age of 16 in one ear, and at the age of 23 in the other. In this case it is probable that the presence of adenoid hypertrophy in the nasopharynx was the factor which determined the earlier onset of the disease, for the remains of the adenoids were still present at the age of 30 when she was first seen by me. The remaining deaf member of the family became affected at the age of 11, but in his case the acute middle ear affection, in all probability, was the exciting cause of the chronic deafness which followed. It may seem strange to associate acute middle ear inflammation with otosclerosis, but as this subject will be referred to later, it need not be discussed at present.

In conclusion, it may be added that, of this family of nine, only one member, D 7, is married. She has had two children, but the older of these is only 6 or 7 years of age, so that it is too early to say whether they are destined to escape the deafness from which their mother and so many uncles and aunts suffer.

The second family tree (Table III) shows some striking differences from, and yet some curious similarities to, the first. It is another example in which a marriage took place between members of two families, each of whom ultimately proved to be burdened with otosclerosis.

Taking the present generation, C, there are six members of the family, and of these three are deaf and have all been examined by me. All are typical cases of otosclerosis.

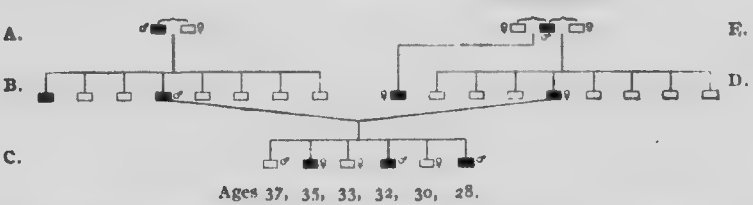
C 1. Male, æt. 37. Does not suffer from deafness or any aural trouble, nor has he at any time in his life had any defect in his hearing.

C 2. Female, æt. 35. The results of examination are tabulated as follows:

Right Ear.		Left Ear.
− 18.	Rinne.	− 15.
+ 7.	Schwabach.	+ 5.
4 ins.	Watch.	3 ins.
1½ yards.	Whisper.	2 ft.
4½ yards.	Conv. voice.	2 yards.
Normal.	Galton's whistle.	Normal.
Lost below mi_2.	Low notes.	Lost below sol_4.

On inspection, it was found that both membranes were quite normal. On inflation through the catheter, air passed freely up the Eustachian tubes, but there resulted little, if any, improvement in the hearing, and the tinnitus was not affected. Tinnitus is constantly present and is surging in character; it made its first appearance four years after the onset of the deafness in the left ear. More recently tinnitus of a booming character has been heard in the right ear. Paracusis is marked. On very rare occasions slight attacks of giddiness have been observed. These have occurred not oftener than once a year, but have been associated with a temporary increase in the tinnitus. The labyrinthine reflexes show a normal response to the caloric tests.

TABLE III



The deafness first appeared at the age of 25; the patient was at that time distinctly anæmic, and was breathless on exertion. On treating the anæmia, the general condition improved very much, and the deafness made no further advance until the age of 30. Since then there has been a slow increase both in the deafness and in the intensity of the tinnitus.

C 3. Female, æt. 33. No deafness or any other trouble associated with the ear is found in this subject.

C 4. Male, æt. 32. The results of examination are tabulated as follows:

Right Ear.		Left Ear.
Normal.	Rinne.	— 10.
Normal.	Schwabach.	—
Normal.	Watch.	Only heard on contact.
Normal.	Whisper.	2 ft.
Normal.	Conv. voice.	4½ yards.
Normal.	Galton's whistle.	Normal.
Normal.	Low notes.	A few lost at lower end of scale.

On inspection, both membranes are found to be normal. Tinnitus and paracusis are both absent, the reason for the absence of the latter being, of course, the fact that one ear has full normal hearing power. The Eustachian tubes are both free, and no noticeable improvement follows inflation.

The dulness of hearing began very gradually and without known cause between the ages of 29 and 30.

The chief interest in this case lies in the fact that at present only one ear is affected. It is very doubtful, however, if this comparatively fortunate condition will last.[1] It is also interesting to note that paracusis is absent at present because the right ear still has normal hearing power. Should deafness of more than a trivial degree occur in that ear, paracusis will probably be noticed.

C 5. Female, æt. 30. The hearing is as yet quite unaffected in either ear. On examination, no defect whatever is found.

C 6. Male, æt. 28. The results of examination are tabulated as follows:

Right Ear.		Left Ear.
— 10.	Rinne.	— 9.
—	Schwabach.	—
3 ins.	Watch.	9 ins.
9 ins.	Whisper.	3 yards.
2 yards.	Conv. voice.	8 yards.
High notes unaffected.	Galton's whistle.	High notes unaffected.
Lost below la_3.	Low notes.	Lost below ut_1.

Weber's test: fork is heard better in the right ear.

On inspection, the right membrane is seen to be somewhat indrawn and is the seat of a small calcareous deposit. The left membrane is also indrawn, but no calcareous deposit is found. On inflation, the air passes freely up both Eustachian tubes. No improvement at all is produced in the right ear by inflation, while in the left ear there is a very slight and transient improvement.

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[1] Since the above examination was made three years ago, the right ear has become affected.

Tinnitus is occasionally present, is slight in degree, and is felt to be in the head rather than in the ears. Paracusis is not observed, the hearing in the left ear being still too good for the occurrence of this symptom.

The history of this case is the usual one in otosclerosis. The dulness of hearing began in the right ear at the age of 22, and in the left at the age of 24. The onset in both ears was very gradual, and without known cause.

In this family, therefore, the normal age onset for otosclerosis appears to be between the ages of 25 and 30. Thus in C 4, where no exciting cause could be discovered, the left ear became affected in the thirtieth year. In C 2 the dulness of hearing began at the age of 25, but for some time previous to that the patient had suffered from chlorosis, and when this condition was rectified the progress of the otosclerosis ceased until the age of 30 was reached. Then the deafness began to increase and has since been progressive.

In C 6 the deafness showed itself in the right ear at the age of 22, and in the left at 24; but this rather earlier onset may well be accounted for by the fact that both membranes were somewhat indrawn, a circumstance indicating some previous catarrhal condition in the Eustachian tube or middle ear. As will be shown later, such factors are of great importance in ante-dating the onset of otosclerosis.

Passing back to the previous generation of the present family, it will be seen that the families B and D of both parents were burdened with deafness, and indeed both parents were themselves deaf. Taking the father's family first, two deaf subjects are found, B 1 and B 4, out of a family of eight. In B 1 the deafness made its appearance between the ages of 50 and 60, and in B 4, the father of the present generation, about the age of 55. Going back still another generation, A, on the paternal side, records can only be obtained of two grandparents. Of these the paternal grandfather became slightly deaf between the ages of 55 and 60, and the paternal grandmother was not deaf at all when she died at the age of 71.

Taking the maternal side of the present generation, the mother herself, D 4, was the only deaf one out of a family of eight, and in her case the deafness has always been and still is comparatively slight. It began at the age of 30 ; but it must be added that the mother had a step-sister, on the maternal grandfather's side, who was deaf. Going back still further on the maternal side, E, only the maternal grandfather and grandmother can be referred to in respect to their hearing ; there is no record of the other members of the family. The maternal grandfather began to be deaf between fifty-five and sixty, and the dulness progressed until he was very deaf when he died at eighty. The maternal grandmother was not deaf when she died, at thirty-five.

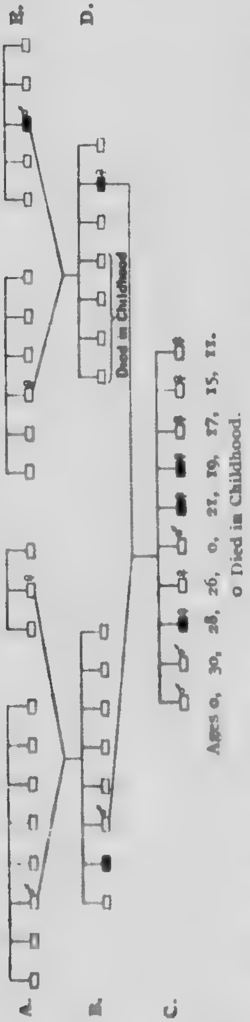
There are in all about forty first cousins of the present generation, and of these only one is deaf. Of second cousins, total number unknown, only two are deaf, but it is a significant fact that these two are brothers—they are on the paternal side.

This family tree, therefore, illustrates again the case in which a marriage takes place between members of two families both of which are only slightly afflicted with deafness. The family resulting from this union, however, is heavily burdened with otosclerosis. It also illustrates again the liability, in such circumstances, for the disease to make its appearance at an earlier age than it did in the parents.

In this connection it is interesting to note that the deafness occurred *after* marriage in both parents, viz. at 55 and 30 respectively.

The third genealogical tree (Table IV) illustrates the case of a family of ten members, but of these, only eight lived to the age at which otosclerosis usually makes its appearance. The inheritance of otosclerosis in this case is, for the most part at any rate, and possibly altogether, from the mother's side. At present only three members of the youngest generation are affected, but it must be observed that the oldest living member is only 30, and there is, therefore, a considerable possibility that others may become deaf. Of the three members affected, all have been examined by me. Regarding the two oldest of these, the diagnosis of otosclerosis is clearly certain, but in respect to the third, some aurists might consider the case doubtful. For reasons to be given later, my own opinion is that it is a case in which otosclerosis was induced at a comparatively early age by a suppurative process in both ears.

TABLE IV



Taking the family of the present generation, C, the following analysis is made :

C 1. Male. Died in childhood before otosclerosis usually manifests itself.

C 2. Male. No defect of hearing.

C 3. Female, æt. 28. The results of examination are tabulated below:

Right Ear.		Left Ear.
− 6.	Rinne.	− 15.
+ 10.	Schwabach.	+ 7.
1 ft.	Watch.	Contact only.
3 yards.	Whisper.	1 in.
6 yards.	Conv. voice.	1 ft.
Mdh. 6·0, Pfl. 2·9.	Galton's whistle.	Mdh. 6·0, Pfl. 4·7.
All heard.	Low notes.	Lost below re$_1$.

Weber's experiment shows that the tuning-fork held in the middle of the forehead is heard better in the left ear than in the right. On inspection of the membranes these appear to be practically normal, but there may be a slight indrawing. There is no rosy tint over the region of the promontory. The Eustachian tubes are normally patent, and after inflation there is no perceptible improvement in either ear. Tinnitus is not present nor is paracusis.

The history of the case is similar to that usually found in otosclerosis. There has never been any earache or discharge from the ear, nor any history of nasal or pharyngeal trouble, and there is no evidence of any previous existence of adenoids in the naso-pharynx. The dulness of hearing began, with gradual onset, in the left ear at the age of 22, and in the right ear at 25.

C 4. Female, æt. 26. There is no defect of hearing so far.

C 5. Male. Died in childhood.

C 6. Female, æt. 21. The results of examination are tabulated below:

Right Ear.		Left Ear.
+ 5.	Rinne.	− 3 or 4
+ 3.	Schwabach.	+ 4.
2 yards.	Watch.	3 in.
5 yards.	Whisper.	1 ft.
8 yards.	Conv. voice.	2 yards.
Normal.	Galton's whistle.	There is a very slight loss for the highest notes
All notes heard.	Low notes.	Lost below la$_1$ ♯.

On applying Weber's test, the sound of the fork is heard better in the left than in the right ear. Both tympanic membranes are normal in appearance and in mobility, and there is no rosy tint present over the promontory. The Eustachian tubes are both perfectly free, and on inflation there is a slight but very transient improvement in hearing. Tinnitus is present in a slight degree, and is rumbling in character. Paracusis is not a symptom in this case

The history of this case is similar to that of the sister previously described. The dulness of hearing began quite recently in the left ear at the age of 21, very gradually and without any discoverable cause. The tinnitus was first noticed a few months after the onset of the deafness. Although the right ear is affected, the deafness is as yet so insignificant that the patient is unaware of it. Indeed the case is following exactly the same course as that of the older sister. There is no history of earache or otorrhœa, and the respiratory passages are, and always have been, quite free of any defect.

C 7. Female, æt. 19. The results of examination are tabulated below :

Right Ear.		Left Ear.
— 5.	Rinne.	— 7.
— 3 or — 4.	Schwabach.	— 5.
Not heard even on contact.	Watch.	Not heard even on contact.
Not heard at all.	Whisper.	Not heard at all.
8 ins.	Loud conv. voice.	4 in.
Mdh. 6·0, Pfl. 3·0.	Galton's whistle.	Mdh. 6·0, Pfl. 4·6.
Lost below ut$_1$.	Low notes.	Lost below ut$_1$.

In response to Weber's test, the patient is unable to say in which ear the fork is heard the better.

On inspection of the right ear, it is found that the membrane is almost completely destroyed, and the tympanic opening of the Eustachian tube is just visible.

All the ossicles are present, and the tip of the hammer is adherent to the promontory; Shrapnell's membrane is not perforated. The stapes and the long process of the anvil are clearly visible, as well as the niche of the oval window, and there are no adhesions in these parts. The round window is very clearly seen, and appears to be quite free from any obstruction. The mucous membrane of the tympanum is thin, dry, and glazed, and there is no cholesteatomatous or epithelial débris.

In the left ear there is a slight amount of purulent discharge, and when this is removed it is found that the appearances are somewhat different from those in the right ear. The tympanic membrane proper has almost entirely disappeared, but the hammer is still present, and at the tip it is adherent to the promontory. There is a perforation in Shrapnell's membrane. The anvil may be present, but it is not visible, neither can the stapes nor oval window be seen, owing to the rather swollen condition of the mucous membrane in this region. The round window is visible, and there is no obstruction in that part.

Paracusis is not noticed, and tinnitus is only occasionally present and is not very marked

The history of the case shows that the patient suffered from double suppurative otitis media at the age of nine. In the right ear this condition disappeared after lasting for about a year, but on the left side it has continued ever since. The patient speaks in the curious monotonous voice of those who become very deaf early in life, but not sufficiently early to cause deaf-mutism. At no time has there been giddiness or any symptom of disease of the vestibule or canals.

As the suppurative process in the left ear had previously resisted all attempts at cure, I performed the radical mastoid operation on Nov 16th, 1910. As a result the suppuration ceased and the ear was quite dry on Jan. 20th, 1911. The hearing was unaffected either favourably or otherwise as a result of the operation.

This case is related rather fully because it serves to illustrate a subject which will be discussed in the section on etiology. At first sight it might have been put down as deafness due to suppurative disease of both middle ears, and such a view, in one sense of the term, would undoubtedly be correct. But a little consideration will show that in this case we are dealing with something more than dulness of hearing due to adhesive processes resulting from the previous otorrhœa. In the first place the only adhesion visible is that binding the hammer to the promontory, and such an adhesion could never cause the profound deafness to which this patient is subject. Further, it is not uncommon to find cases in which changes are present almost identical with those just described, and yet the degree of deafness is usually comparatively slight. But another even more cogent reason which prevents us considering the deafness in this case to be entirely the result of changes in the middle ear is the fact that the bone-conduction is noticeably diminished. It is true that Rinne's test is negative, but this is only due to the fact that the air-conduction is even more seriously affected than the bone-conduction. Schwabach's test shows that the bone-conduction is diminished by three or four seconds on the right side and by five seconds on the left.

The absence of paracusis is also, to a certain extent, indicative of something more than uncomplicated middle ear disease.

While, therefore, we cannot definitely state that in this case otosclerosis is present, we are justified in assuming that it probably is, especially when taken in consideration with case D 1 in Table II and with case

B 1 in Table VI to be described later. In both these cases otosclerosis is unquestionably present, though it began immediately after acute middle ear inflammation.

Finally, in the case under discussion, otosclerosis might very naturally be expected since two of the older sisters suffer from it; and deafness was present in the mother, and is also found in an aunt on the father's side.

Taking the next generation back on the paternal side, B, it is seen that the father himself is not affected, and of the whole family of eight only one is dull of hearing. The degree of inheritance on this side, so far as deafness is concerned, is very slight, if indeed there be any at all.

The next generation back on the paternal side shows that there is no discoverable history of deafness either in the family of the paternal grandfather or the paternal grandmother.

Of the great-grandparents on the paternal side nothing is recorded in respect to the hearing.

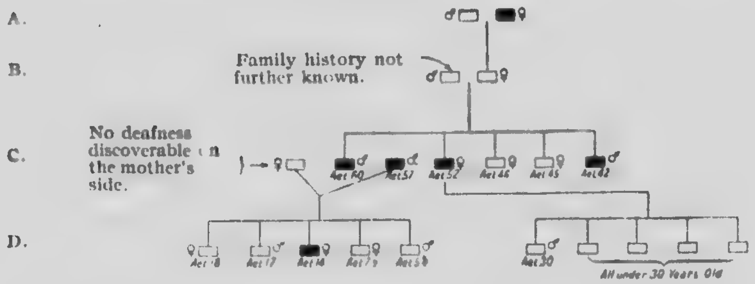
Taking the maternal side, the mother herself, D 6, was deaf; and from the description of her symptoms given by her children, it is very probable that otosclerosis was the cause of the deafness. She suffered from dulness of hearing, tinnitus and paracusis. The mother's own family consisted of seven members, but of these only three reached the age at which otosclerosis is liable to occur, and the family, therefore, must be considered as having produced one sufferer out of three.

In the preceding generation, the maternal grandmother's own family consisted of six, who were all unaffected by deafness. The maternal grandfather's family also consisted of six, of whom the grandfather himself, E 3, was the only deaf one. The maternal great-grandmother was deaf when old, but it is not known how long she suffered.

This family tree, therefore, illustrates the case in which the tendency to otosclerosis is inherited for at least the most part and perhaps entirely, from the maternal side. This is said to be commoner than inheritance from the paternal side.

The next family tree (Table V) illustrates the case in

TABLE V



which, so far as is known, the tendency was inherited entirely from the paternal side.

The present generation is young, the oldest being 18, and the youngest 5½ years of age. The family consists of five members.

D 1. Female, æt. 18. Hearing unaffected.

D 2. Male, æt. 17. Hearing unaffected.

D 3. Female, æt. 14. Subject of otosclerosis. The results of the tests for hearing are as follows:

Right Ear.		Left Ear.
− 10.	Rinne.	− 12.
+ 8.	Schwabach.	+ 10.
2 ft.	Watch.	1½ ft.
4 yards.	Whisper.	1 yard.
8 yards +.	Conv. voice.	8 yards +.
All notes heard up to normal.	Galton's whistle.	All notes heard up to normal.
A few notes lost at lower end of scale.	Low notes.	A few notes lost at lower end of scale.

On inspection of the tympanic membranes both are found to present perfectly normal appearances. No rosy tint is seen over the region of the promontory. The mobility of the membranes is normal on both sides.

On inflation with the catheter air passes freely up the Eustachian tubes of both sides, but no appreciable improvement results.

Tinnitus of a hissing character is present, but paracusis has not yet been observed, since the dulness of hearing is not yet sufficiently pronounced. There is, however, considerable probability that paracusis will appear as the deafness increases.

The history of the case reveals the following facts. The dulness of hearing was first noticed about a year before the examination. It was of gradual onset and there was never any pain or discharge of any kind. The patient first noticed the tinnitus a few months after the dulness of hearing was suspected.

This is a case in which otosclerosis has made its appearance at a relatively early age, especially in view of the fact that there is no obvious exciting cause, such as was found in case D 1 of Table I, and in case C 7 of Table IV.

D 4. Female, æt. 7½. There is no affection of the organ of hearing.

D 5. Male, æt. 5½. There is no affection of the organ of hearing.

In this family, therefore, although none of the members has yet reached the usual age for the appearance of otosclerosis, one of them already manifests the condition unmistakably. Probably one or more of the remaining members will ultimately become sufferers.

Taking the next generation back on the paternal side, it will be seen that the father's own family consists of six members. Of these four are deaf. The analysis is as follows:

C 1. Male, æt. 60. Not examined by myself. The patient first showed signs of deafness at the age of 40, and the symptom has gradually increased ever since. He suffers also from tinnitus and paracusis. There has never been any suppuration, earache, or history of nasal or naso-pharyngeal trouble.

C 2. Male, æt. 57. Father of the family described above. The following table shows the results of the tests for hearing :

Right Ear.		Left Ear.
− 10.	Rinne.	− 12.
+ 6.	Schwabach.	+ 9 or + 10.
Not heard at all.	Watch.	Not heard at all.
Not heard at all.	Whisper.	Not heard at all.
10 ins.	Conv. voice.	1 ft.
Lost above Mdh. 8·0, Pfl. 10·4.	Galton's whistle.	Lost above Mdh. 8·0, Pfl. 10·8.
Lost below re_3.	Low notes.	Lost below re_3.

On inspection, both tympanic membranes present normal appearances. No rosy tint is seen over the region of the promontory. The Eustachian tubes are perfectly free, and on inflation air passes quite unobstructed into the tympanum. There is, however, no improvement whatever after inflation. There is no history of giddiness or any symptoms pointing to disease of the vestibule or semi-circular canals, and the caloric nystagmus tests indicate a normal condition of these structures. Tinnitus is not complained of at all at present. The patient, however, remembers that at one time, about fifteen or twenty years ago, he noticed it for several months or perhaps a year, but the symptom passed away and was not very distressing. Paracusis is very marked, and has been so for at least fifteen or twenty years.

The deafness began very gradually and without known cause about the age of 31. There was no pain or discharge at any time. Both ears were affected at approximately the same time, as far as the patient can remember. In recent years the deafness has tended to become distinctly worse, and it is quite possible that this is to be associated with degenerative changes in the sound-perceiving apparatus. It is true that the bone-conduction for the low notes is still above normal, but it will be observed that the hearing power for the high notes is seriously affected.

C 3. Female, æt. 52. Not examined by myself.

The patient first showed deafness about the age of 30. Tinnitus and paracusis are both present. The patient is married and has five children, none of whom is as yet deaf, but it must be pointed out that the oldest of these is only 30, and it is very possible that otosclerosis may make its appearance in some members of the family later.

C 4. Female, æt. 46. Unmarried. No affection of the hearing.

C 5. Female, æt. 45. Unmarried. No affection of the hearing.

C 6. Male, æt. 42. Not examined by myself. The deafness, which in this case is associated with tinnitus, only began a year ago, and so far is confined to one ear. The onset was gradual and without pain or known cause.

Taking the next generation back, B, the only record that can be obtained is that neither parent was deaf although both lived until over 60. Nothing is known of the brothers and sisters in this generation.

Going back still another generation, A, to the great-grandparents of the present young generation, all that is known is that one member of the family was very deaf at the age of 70, and probably for a considerable time before. This was the great-grandmother of the youngest generation.

On the maternal side of the present younger generation there is no history of deafness discoverable even on careful examination.

One of the points of interest in this family tree therefore, lies, first in the very early age at which otosclerosis has made its appearance in the present younger generation. Another interesting feature is the fact that the present older generation is heavily burdened with otosclerosis (four out of six), in spite of the fact that neither of the parents was deaf.

The next family tree (Table VI) is, unfortunately, at present very incomplete. The patient states, however, that deafness has long been recognised as being in the family on the maternal side. There is no discoverable family history of deafness on the father's side.

Taking the present generation, B, the family is seen to consist of six members, of whom the oldest is 41, and the youngest 21 years of age. Of the six, three are deaf; and of these only one has been examined by myself. The family medical attendant, however, informed me that the other two deaf individuals have been examined by aurists, and have been definitely certified to be suffering from otosclerosis.

B 1. Female, æt. 41. Examined by myself. The results of the tests for hearing are given below:

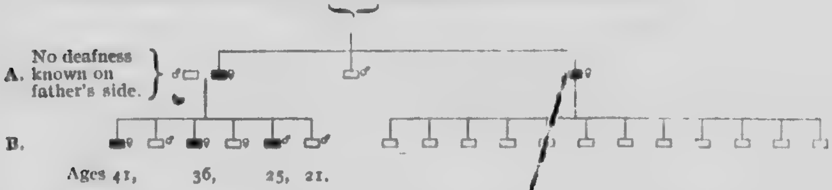
Right Ear.		Left Ear.
− 10.	Rinne.	− 15.
+ 8.	Schwabach.	+ 9.
Not heard even on contact.	Watch.	Not heard even on contact.
2 yards.	Whisper.	6 ins.
5 yards.	Conv. voice.	1 yard.
High notes all heard.	Galton's whistle.	Lost above Mdh 6·0, Pfl. 4·9.
Lost below fa_1.	Low notes.	Lost below si_1.
Weber to left.		

On inspection of the right membrane it is found that there is neither indrawing nor any thickening or atrophy or other change in the membrane itself. The long process of the anvil is visible. There is a very distinct rosy tint over the promontory. The left membrane is indrawn, but there is no thickening or atrophy. A distinct rosy tint is observed over the promontory in exactly the same position as that on the right side. On inflation by means of the catheter, air passes freely into both tympana, and there is no obstruction in the Eustachian tubes. There is no improvement in hearing after inflation.

Paracusis has not yet been observed. Tinnitus has never been present in the right ear at all. In the left ear it was present for several years in a mild degree, but during the last six months it has not been noticed.

TABLE VI

Mother's ancestors known to have been severely affected with deafness.



Now the case just described is a typical example of otosclerosis. But the interest begins when the history of the aural condition is studied in detail.

Until sixteen years ago, when the patient was 25, the hearing was perfectly good. At that time she was out hunting one day and had a long run with a very cold east wind blowing on the left side. An hour after reaching home she began to suffer from acute pain in the left ear, and noticed that she was deaf. The pain in the ear lasted for nearly a week and the deafness increased. The pain then passed away and there was no discharge from the ear at any time. The deafness, however, remained and has never improved, in spite of the fact that she had the attention of several highly skilled aurists. Tinnitus made its appearance in the left ear a number of years later, but was not very severe.

The right ear remained perfectly well until three years ago, when she began to notice that it also was gradually becoming dull of hearing. This was associated with tinnitus of a mild degree. A few months previous to this the patient suffered from a slight attack of appendicitis, but it did not occur to the patient herself or to her medical attendant that there might be any association between the abdominal trouble and the dulness of hearing. The hearing gradually became worse, and the patient was treated for this, but without obtaining any relief. A second attack of appendicitis occurred from which she again recovered without operation. A third and much more severe attack occurred in May 1910, and this necessitated operation for removal of the appendix. She made an excellent recovery and has been free from abdominal trouble since. About three or four months after the operation the patient noticed that the tinnitus had disappeared, but she did not associate this with the removal of the appendix. Indeed the relationship, if any, which existed between the abdominal trouble and the otosclerosis was not suggested until I saw her in December 1910.

This case is described in detail because it illustrates in a remarkable manner the fact that either some general constitutional condition, such as a toxin or other substance in the blood, or a local inflammatory condition in the middle ear may be the exciting cause of otosclerosis.

In this particular case the unique feature is that in the left ear the exciting factor was undoubtedly the middle ear inflammation, and in the right ear it was very probably the toxin or toxins absorbed from the diseased appendix. It is, of course, quite possible that the diseased appendix may have had no relationship to the onset of the otosclerosis. In any case, however, the exciting cause of the otosclerosis in the right ear was not a middle ear inflammation, as it was in the left; nor was there any evidence that it was due to any middle ear condition at all. The cessation of the tinnitus after the removal of the appendix indicates rather that the latter was the exciting cause of the otosclerosis in the right ear.

But the case illustrates another point quite as important as that just discussed. In this family the inherited tendency to otosclerosis is so marked as to be unmistakable, and the real relationship of the acute middle ear inflammation to the otosclerosis is seen to be that merely of the exciting cause. Let it be supposed, however, that the inherited tendency, though present, had been less pronounced, and perhaps not discoverable; then the condition in the left ear might very naturally have been ascribed to acute middle ear inflammation without any qualifying statement as to heredity. I have little doubt that such incompleteness in diagnosis must occasionally occur in the practice of all aurists; I am sure it must have done so in mine.

B 2. Male. There is no affection of hearing.

B 3. Female, æt. 36. This patient was not examined by myself, but has been seen by various aurists, and diagnosed as "chronic dry catarrh" and as "otosclerosis." The dulness of hearing began at the age of 20 and is accompanied by tinnitus and paracusis.

B 4. Female. Hearing not affected in any way.

B 5. Male, æt. 25. Not examined by myself. The patient is deaf and suffers from tinnitus. He had been delicate for several years when the dulness of hearing began at the age of 19; and his medical attendant informed me that he had seen at least two aurists, both of whom diagnosed the condition as otosclerosis.

B 6. Male, æt. 21. There is no affection of hearing.

As regards the families (A) of the parents of the present generation, no dulness of hearing is recorded on the father's side at all. As regards the maternal side, it is seen that out of a family of three, two are deaf, and of these the mother herself is one. From the family medical attendant I have learnt that the mother's case was diagnosed as one of "chronic dry catarrh," a term which at that time included otosclerosis. Concerning the other deaf member of this family no record can be obtained at present.

As stated above, this is a family in which the individual members recognise clearly that the tendency to deafness has been present for several generations, but I am not at present able to give any further data concerning the family tree.

The next family tree (Table VII) illustrates the case in which the hereditary element does not appear to be markedly present; indeed we have no real proof that heredity has played any part in the occurrence of the disease. For my own part, however, I have no doubt as to the hereditary influence in this case. The occurrence of otosclerosis in four members out of a family of eleven, associated with the fact that the age incidences were fairly similar, 19, 24, 26, 29, and that no exciting cause, local or constitutional, could be found, is in itself, in my opinion, sufficient ground for drawing the inference that heredity has played a part in the condition, apart altogether from the fact that in this case the grandmother became deaf some time before the age of 50.

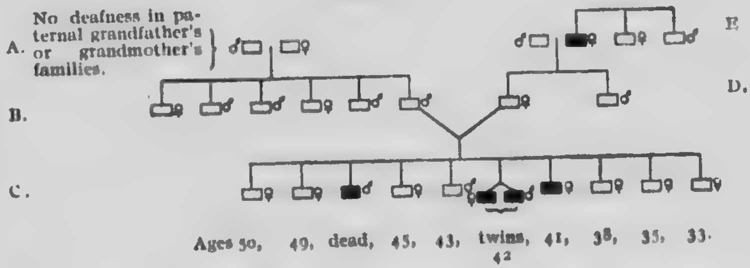
The following is the analysis of the tree, so far as I have been able to carry it out:

C 1. Female, æt. 50. Not deaf.

C 2. Female, æt. 49. Not deaf.

C 3. Male; dead. Began to grow deaf at the age of 26 without any known cause. He suffered from tinnitus and paracusis, and the case was diagnosed by an aurist and by his own medical attendant as "chronic dry catarrh," a term which at that time included otosclerosis.

TABLE VII


No deafness in paternal grandfather's or grandmother's families.
A.
B.
C.
D.
E
Ages 50, 49, dead, 45, 43, twins, 42, 41, 38, 35, 33.

C 4. Female, æt. 45. Not deaf.

C 5. Male, æt. 43. Not deaf.

C 6. Female, and twin with C 7, æt. 42. Deafness began at the age of 29, and is associated with marked tinnitus.

C 7. Male and twin with C 6, æt. 42. Began to be deaf at the age of 19 or 20, suffers from tinnitus, and is the subject of paracusis.

C 8. Female, æt. 41. Examined by myself. The results of the tests are tabulated below:

Right Ear.		Left Ear.
− 15.	Rinne.	− 10.
+ 4.	Schwabach.	+ 5.
Not heard even on contact.	Watch.	Not heard even on contact.
Not heard at all.	Whisper.	Not heard at all.
6 ins.	Conv. voice.	7 ins.
.	Low notes.	Lost below la$_3$ 2.
Lost above Mdh. 7'0, Pfl. 10'3.	Galton's whistle.	Lost above Mdh. 7'0, Pfl. 5'8

On inspection it is found that the right tympanic membrane is quite normal in appearance in every respect, and there is no rosy tint over the region of the promontory. In the left membrane there is a slight deposit of calcareous salts in the anterior quadrants, but there is no indrawing of the membrane nor is any rosy tint present over the region of the promontory.

On inflation through the catheter air enters the tympanum freely on both sides, and there is no obstruction in the Eustachian tubes. No improvement resulted in either ear after inflation.

The history of the case is similar to that usually recorded in otosclerosis. The dulness of hearing was first noticed about the age of 24 or 25. No cause could be found for the dulness of hearing. The patient has never suffered from any noticeable tendency to nasal or nasopharyngeal trouble, and no history of anæmia is recorded. Paracusis is present. At no time was there any giddiness, and on employing the caloric tests the vestibular reflexes are found to be normal.

C 9. Female, æt. 38. No affection of hearing.

C 10. Female, æt. 35. No affection of hearing.

C 11. Female, æt. 33. No affection of hearing.

Taking the next generation (B) back on the paternal side, it will be seen that neither the father, who died at 70, nor any of the members of his own family suffered from deafness. In the next generation (A) on the paternal side, it is found that neither of the grandparents was deaf, nor is there any record of deafness among the grand-aunts or grand-uncles.

On the maternal side, the mother's own family (D) consisted of two, including the mother herself, who died at 74. Neither of the two was deaf. In the next generation back (E) the grandmother's own family consisted of three individuals, of whom only the grandmother herself was deaf. She became dull of hearing at the age of fifty, and probably for some little time before then. The nature of the deafness cannot be ascertained. Of the maternal grandfather's family nothing is known, beyond the fact that he himself was not deaf even in comparatively old age.

There are two points of interest in this family tree. The first is the fact that two of the victims of otosclerosis were twins. I am not aware of any other recorded case similar to this, but it is quite possible that such a coincidence has been noted. The second point of interest is the meagreness of the direct evidence of the hereditary element. Indeed, as previously stated, the deafness in the grandmother of the present generation may not have been due to otosclerosis.

From indirect evidence, however, I think there can be no doubt that the hereditary tendency is present. When four individuals out of a family of eleven develop the affection without any discoverable cause, we may fairly safely assume, from what we know of the manifestation of the disease in families where the hereditary influence is obvious, that in this case also the same cause is at work, though we may not be able to trace it by direct inspection of the family tree.

Another point of interest is that, assuming the grandmother to have been the victim of otosclerosis, the intervening generation (D) has escaped. This, however, is not an uncommon occurrence in other conditions besides otosclerosis.

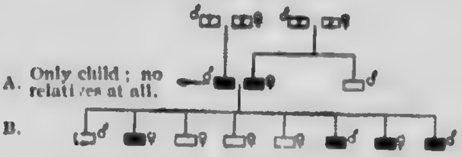
The next family tree (Table VIII) can only be recorded over the present generation and that immediately preceding it. It is given for two purposes. The first of these is to illustrate what is very possibly an example of inherited tendency to otosclerosis from both father and mother. The second purpose is to show how persistently some individuals refuse to admit any hereditary defect in regard to deafness when the facts so strongly point in this direction.

In the present generation (B) there were originally eight members in the family, but of these the oldest died in childhood, that is, before the age at which otosclerosis makes it appearance. Of the remaining seven, four are deaf, but only in one of these have I had the opportunity of making a personal examination.

B 1. Male. Died in childhood.

B 2. Female, æt. 55. Deaf in both ears. The deafness is said to have come on after scarlet fever, but it is bilateral, and there is no known history of suppuration. On the other hand paracusis is marked, and the evidence is just as much in favour of the condition being otosclerosis as adhesive processes in the middle ear the result of scarlet fever. In all probability this is a case in which the scarlet fever merely played the part of the exciting cause, bringing to light the inherited tendency to otosclerosis.

TABLE VIII



B 3. Female, æt. 53. No defect of hearing.

B 4. Female, æt. 53. No defect of hearing.

B 5. Female, æt. 47. No defect of hearing.

B 6. Male, æt. 45. Dulness of hearing in both ears, associated with paracusis and tinnitus. There is no history of suppuration or pain in the ear. The dulness of hearing is attributed to a fall from a horse. On inquiry it appears that the accident was not very serious, for the patient was up and out of doors two days afterwards. The dulness of hearing first made its appearance a few weeks later. There is no history of giddiness or of any symptoms which would point to injury of the auditory nerve or labyrinth.

This is a case in which obviously the injury could not be the sole cause of the dulness of hearing. The real cause, in all probability, is to be found in the hereditary tendency to otosclerosis, though of course the accident may have been the exciting factor in revealing the weakness.

B 7. Female, æt. 41. Examined by myself. The results of the hearing tests are tabulated below :

Right Ear.		Left Ear.
− 12.	Rinne.	+ 6.
—	Schwabach.	—
9 to 10 ins.	Watch.	1 yard.
2 yards.	Whisper.	5 yards.
6 yards.	Conv. voice.	8 yards.
Heard up to normal.	Galton's whistle.	Heard up to normal.
Lost below sol_2.	Low notes.	All heard.
Weber to right.		

On inspection, both membranes are seen to be quite normal ; there is no indrawing, nor any rosy tint over the region of the promontory. On inflation through the catheter, air enters the tympanum freely and no obstruction is present in the Eustachian tubes. There is improvement to a very slight extent after inflation, but it is very fleeting. The nasal and pharyngeal cavities are quite normal. Nasal catarrh and damp weather do not affect the hearing, but exhaustion makes it much worse. The dulness of hearing on the left side is, at present, so slight that the patient is unaware that there is any, but as will be seen from the table, the watch is only heard at about half the normal hearing distance. Tinnitus of a singing character and very slight in degree made its appearance a few weeks ago. Paracusis has never been present; the hearing in the left ear being too good to permit of the occurrence of this symptom. There has never been any symptom pointing to disease of the canals or vestibule.

The only cause for the deafness of which the patient can think is a rather lengthened period of sea-sickness. Such a condition is, after all, almost as unlikely to be the exciting cause of otosclerosis as the fall from the horse, in the case of her brother. The real essential cause was probably the inherited tendency.

B 8. Male, æt. 38. Not examined by myself.

The deafness is bilateral, is considerable in degree, and is associated with paracusis but not with tinnitus. The condition has been attributed in this case to neglected colds and polypus in the nose. In this case also the nasal trouble was only the exciting cause.

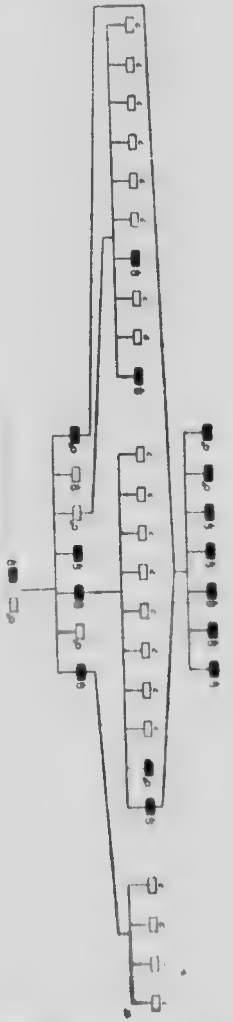
Taking the next generation back (A) on the paternal side, the family consisted of one member only, the father himself. He suffered from deafness of considerable degree and associated with paracusis. It is not known whether tinnitus was present or not. The dulness of hearing came on gradually in early middle life. Nothing is known of the grandparents on the paternal side.

On the maternal side, the mother's family consisted of two, the mother herself, C 1, and a brother. Of these the mother suffered from bilateral deafness associated with paracusis, but whether tinnitus was present or not cannot now be discovered. The uncle, C 2, is not deaf. Nothing is known of the grandparents on the mother's side.

In this family tree we have, therefore, a case illustrating an inherited tendency to otosclerosis, but we cannot say whether the tendency was derived from the mother's side or from the father's, but there is fair probability that both contributed. It is to be noted that among the members of this family it is firmly believed that there is no hereditary tendency to deafness. The case among many others illustrates the necessity of examining closely into the details of the family history, and never accepting the bare statement of patients that there is no hereditary tendency to deafness in their family.

In addition to these family trees which I have described in the previous pages, I take the liberty of including one which was investigated by Hammerschlag and referred to by Körner. It illustrates the unique case of a man, the victim of otosclerosis, marrying his niece who also suffered from the same condition. There were seven offspring from this marriage, and all of the seven became deaf. This is, so far as I am aware, the most striking example of the effect of inherited tendency in the production of otosclerosis. It is not, however, perhaps so remarkable in some ways as the case shown in Table II, in which six members of a family of nine are the subjects of otosclerosis, and in which very possibly eight out of the nine may become affected, although both parents were free of the disease.

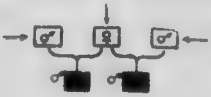
TABLE IX
(From Hammerschlag)



Finally, an interesting case of otosclerosis occurring in members of the same family is recorded by Paul-Boncour. The family tree is not given in the original paper, but the writer evidently made a very careful investigation into the family history of the mother, and an equally careful examination of the two sufferers, who were half-brothers. Both patients began to suffer from typical otosclerosis without any obvious exciting cause, at an age relatively very young, 14 and 12 years respectively. The mother was married twice, and to each husband she bore a son, the patients just described.

TABLE X

No discoverable history of deafness in the mother's family.



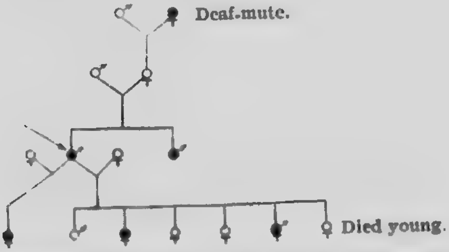
The family history of the first husband not given.

The family history of the second husband not given.

The mother herself was not deaf, nor, on the closest examination, could any history of deafness be found in her family. It is unfortunate that no records of the family histories of the husbands are given in this case, because absence of such data leaves several possibilities open as explanations. Thus, it cannot be definitely stated that the tendency to otosclerosis has been inherited from the mother, for it is quite possible that, by coincidence both husbands, though not deaf themselves, may have come of a stock in which deafness was hereditary. Thus we are left in considerable doubt as to the extent which heredity has played in this case. It is interesting, however, as showing how otosclerosis may manifest itself in a typical and severe manner, even when there is no evidence of the condition having been present in any of the ancestors.

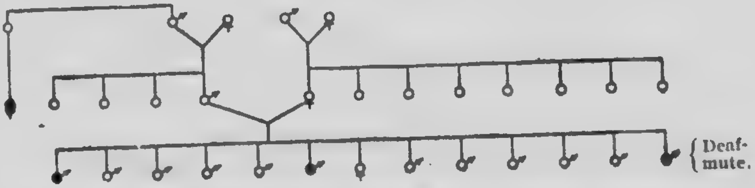
In addition to the family trees of which the detailed investigation is given above, several others are shown below with notes indicating the chief points of interest.

TABLE XI



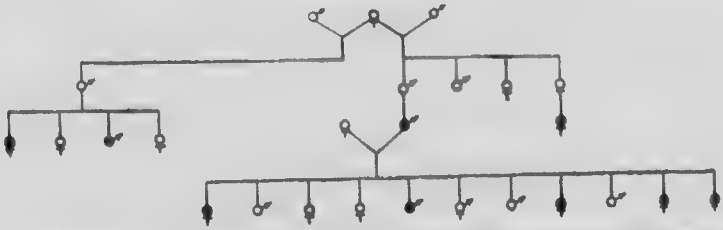
In this tree the great-grandmother was deaf-mute, but it is not possible to discover whether the affection was acquired or inherited; and no inference therefore can be drawn as to whether the otosclerosis which appeared later in the family tree was associated with the deaf-mutism or not. In the grandparents there was no deafness, but both their children suffered from otosclerosis, and the elder of these was married twice and had two families, in both of which otosclerosis occurred.

TABLE XII



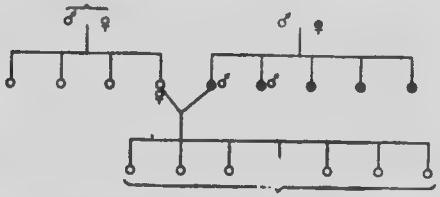
In this large family otosclerosis made its appearance in two members, and deaf-mutism in a third.

TABLE XIII



In this family tree the great-grandmother was married twice, and neither of her husbands nor herself was deaf. She had a family by each husband, but no deafness appeared in either of these families. The children of each of these families were, however, burdened with otosclerosis, and the next generation of one of the subjects had eleven children, of whom five became the subjects of the disease.

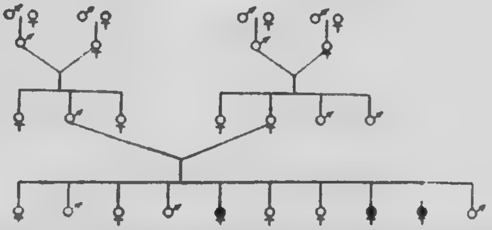
TABLE XIV



None deaf, although all have reached the age at which otosclerosis usually manifests itself.

This table shows the case of one deaf parent who had five children, all of whom fell victims to otosclerosis—a very severe incidence. On the other hand, one of these patients had seven children, none of whom developed otosclerosis, though all have passed the age at which the disease usually occurs.

TABLE XV



This family tree is interesting as showing how otosclerosis in its typical form has made its appearance in those members of a family in which there was no deafness in the parents, the grandparents, nor the great-grandparents.

CHAPTER III

BIOLOGICAL

Now, before going on to discuss more closely the mysterious nature of the origin of otosclerosis, and the part which hereditary tendency plays, it must be clearly pointed out that, as every aurist is aware, not a few cases of undoubted otosclerosis occur in which no hereditary tendency can be discovered. The tables given above are only cited to show, in their detailed relationships, cases in which the hereditary element is fairly obvious. Further, in my own cases, I have gone into considerable individual detail, even at the risk of being prolix, in order to show how easy it is to fall into the error of attributing the occurrence of otosclerosis to other factors than heredity, because they are the ones which the patient always offers to the aurist for consideration first, and frequently alone.

For the same reason it will not be difficult to see why it is impossible to state the percentages of cases of otosclerosis due to heredity, and the number due to other causes. Any such division is purely arbitrary, because in a very large proportion, and perhaps in all cases, inherited tendency and some adventitious cause each play a part. Thus, in the case of Table VI, B 1, it would be impossible to ignore hereditary influence on the one hand and acute middle ear disease and toxic absorption on the other. A similar difficulty arises in case C 7 in Table IV, and in case D 1 in Table II. I had, at one time during this investigation, intended to have a percentage table drawn up, but as the research evolved itself, and individual cases were examined in detail, as exemplified in not a few of those described, it became quite evident that the data which could be offered for statistical treatment were not sufficiently definite for the latter purpose. And it is not surprising, therefore, that the percentage of cases due to inheritance, given by different authorities, should vary within such wide limits. It is doubtful if the hereditary tendency, using the term in its widest sense, can ever be definitely excluded in a given case of otosclerosis, because it can always be maintained that the genealogy is not sufficiently well known. On the other hand, an exciting cause may possibly be acting which is not discoverable by the present methods of physiological and clinical investigation.

There is one point, however, which I venture to say may quite justifiably be considered to be proved. It is this, that apart from hereditary influence there is no single cause to which otosclerosis may be attributed. Now this is an important practical point in considering our mental attitude towards the disease. It is illustrated in several of the cases described in the preceding pages. Particularly striking is the case B 1 in Table VI, in which one ear became affected by otosclerosis as a result of acute middle ear inflammation, while the other was clearly enough the result of another cause, probably the presence of some toxic substance in the blood. In both ears, of course, the hereditary influence played its part. Hence, attempts to ascribe all cases of otosclerosis to disturbances in the internal secretions of [illegible] gland or the pituitary body, toxic absorption from [illegible] alimentary canal or other mucous surface, sy[illegible] cannot be accepted, although in a give[illegible] these may be the exciting cause.

Similarly there is no justification for the view that in all cases the exciting cause is to be found in past or present inflammatory processes in the middle ear. It is undoubtedly true that in not a few cases, middle ear disease is clearly the exciting factor in the production of otosclerosis. This has been already shown clinically in the preceding pages, and in recent times Fraser[1] has demonstrated the fact by direct pathological evidence. But it is just as clear that many cases arise in individuals in whom there is no evidence whatever, either from a clinical or a pathological side, of any affection of the tympanum, past or present.

As regards the incidence of the disease in relation to age and sex, the subject has already been disc[illegible] in previous pages. But there is another aspect of [illegible] question of age incidence which is not discussed [illegible] certain families in which otosclerosis is definitely [illegible]ditary, or to put it more accurately, when the he[illegible] influence [illegible] pronounced the disease appears [illegible] an earlier [illegible] in each successive generation. [illegible] is seen in Tables [illegible] III and V. The number of trees, however, which are available is too few for the pur[illegible]ing any very definite conclusions. The suggestion is made now, in order that any reader who thinks of [illegible] up the subject may be on the alert to m[illegible] in respect to this matter. Before leaving [illegible] subject, I should like to add that Dr. Mott has co[illegible] similar conclusion in respect to certain diseases of the nervous system in which the influence of heredity is particularly noticeable.[2] But Dr. Mott's evidence is drawn from a very much larger number of cases and more definite conclusions can, therefore, be inferred from them. Should it be found after an inspection of a large number of genealogical trees, that the suggestion just made in regarding otosclerosis can be definitely shown to be correct, then a very interesting link will be established in respect to the bearing of hereditary influence on diseases of the central nervous system and otosclerosis.

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[1] Fraser, *Roy. Soc. Med., Otolog. Sect.*, May 1916.

[2] Mott, *Archiv. of Neurology*, Vol. V. Appendix, 1911.

Another inference which may be drawn from a consideration of the facts recorded in the preceding pages, is that the age of onset of otosclerosis may be very greatly affected by local inflammatory activity in the middle ear. This is illustrated in case D 1 in Table II and case C 7 in Table IV. In these subjects there was clearly an inherited tendency to otosclerosis; but whereas in ordinary circumstances the disease does not manifest itself until after the twentieth year, they became affected very early, viz. at the ages of 11 and 9 respectively. This is a matter of great practical importance from two points of view. The first is that the children of parents who are the victims of otosclerosis should be guarded with especial care against every possible factor which may tend, even remotely, to middle ear disease of any kind. Thus the mildest catarrhal conditions in the nose or naso-pharynx or pharynx call for interference more imperatively even than in the children of those in whom no inherited tendency to otosclerosis exists. The second deduction that can be made from the foregoing facts is, that in all cases of deafness, even in children, the family history should be inquired into very carefully, and if there is evidence of a tendency to otosclerosis, the prognosis should be guarded, however much the case may present the clinical signs and symptoms of Eustachian or middle ear catarrh, or acute middle ear inflammation.

It is not sufficient merely to associate the incidence of otosclerosis with hereditary tendency. Many cases come before the aurist in which no hereditary element can be detected, though in these cases of course, as stated above, the disease may have been present in a remote ancestor. Even admitting this, the problem still remains unanswered; because, however far we may go back in the ancestral tree, the question still remains, how did the condition originate? It can hardly be imagined that there is an unbroken descent of actual cases of otosclerosis from the time when the organ of hearing first made its appearance in the animal kingdom. Further, any theory of the etiology of otosclerosis must explain several very well known but mysterious features about the disease, such as the almost universal bilateral nature of the condition, and the remarkable fact that, with very rare exceptions, the vestibule and semicircular canals are unaffected. It must also explain the equally curious fact that in many cases, in the later stages, the nervous apparatus of the cochlea tends to become diseased, whereas that of the vestibule and canals remains unaffected.

It is for these reasons that the writer is unable to accept the views of Körner that otosclerosis is always inherited. Indeed, Körner himself admits the difficulty of this explanation, which, in reality, is no explanation at all. Otosclerosis must have originated some time in the history of the race; and, that being so, is there any reason why it should not occasionally originate in the present time? The condition is even found in animals other than man, as has been shown by Alexander and Katz, and presumably it must have originated in these animals also.

Another fact which must be kept in mind is that no single general or local constitutional condition can be associated with otosclerosis. Indeed it is from the latter fact that a hint may be obtained as to the point at which a search should be made in considering the etiology of the disease.

When the pathologist is confronted with a problem in which an abnormal local condition arises that cannot be accounted for in different cases by the action of a specific micro-organism or a definite toxic product, and when, moreover, there are no signs of inflammatory activity, it behoves him to examine carefully the process of evolution by which the organ and tissues, with which he is dealing, came into existence. There are several problems of this nature before the pathologist at the present time, but apparently this method of examining them has had little, if any, attention paid to it. An example of such a problem is the reason for the relatively great frequency of cancer in certain situations, such as the breast, rectum, and uterus, compared with its incidence in other localities. Another deeply interesting problem is the discovery of the real basis on which rests the occurrence of arterio-sclerosis. Similarly, the frequency with which skin affections show a marked predilection for certain areas is a mystery which requires solution. Among such problems the occurrence of otosclerosis is one, and in this case, I venture to think that the solution is to be found in the evolution of the organ of hearing.

It is, of course, outside the scope of this work to go into a detailed description of the whole organ of hearing, but for the purpose at present in view it is necessary to consider certain aspects of the subject before attempting to explain directly the relationship which the process of evolution bears to otosclerosis.

There is, of course, no possibility of tracing by direct observation the increasing complexity of the organ of hearing in the actual species of vertebrates from which the mammals have been gradually evolved. There is, however, quite a sufficient number of living species to give, by inference, a fairly clear picture of most of these processes, though there are some gaps which cannot at present be bridged.

In the fishes, considerable variations are found in the structure of the labyrinth. If amphioxus be excluded for the present, the simplest form is that of the very primitive hag-fish (*Myxine glutinosa*). In many fishes, on the other hand, the labyrinth is exceedingly complex. Indeed, so far as the organs of the saccule, utricle and semicircular canals are concerned it may be said that the organ never again reaches such a degree of complexity. To the great majority of these it is not necessary to refer, as they are obviously far out of the line of direct ancestry of the other vertebrate classes. Of one of the sub-classes of the fishes, the dipnoi, however, it is necessary to say a few words, because of its remarkably close approximation, in many respects, to the amphibia. Owing to the kindness of Professor Graham Kerr, I have been able to prepare a specimen of the labyrinth of the lepidosiren. Another example of the labyrinth of the dipnoid fishes has been carefully described by Retzius. In both these fishes the semicircular canals and the structures of the vestibule are relatively well developed, quite as highly, that is to say, as in the amphibia, reptile, or mammal. Two points, however, are to be noted. In the first place there is no rudiment of a cochlea, nor even the special macula basilaris from which the cochlea is ultimately to be evolved. The next point, which I have verified by dissection of the organ in the lepidosiren, is that there is no trace of any opening in the labyrinth corresponding to the oval or round windows, nor any chain of ossicles or tympanic membrane.

In the amphibians there is considerable variety in the structure and connections of the labyrinth. In the more highly specialised, such as the frog, a tympanic membrane is present, and it is in direct connection with the labyrinth by means of a columella, similar to that found in reptiles and birds. The relationship of the inner end of the columella to the labyrinth is much more complex, however, than is the case in the reptile and in the bird. But there is no necessity to enter into this at present, because the more highly specialised amphibia of the present day cannot be looked upon as in the direct line of ancestry of the reptile or mammal.

In respect to the less specialised amphibia which have a more direct bearing on the subject under discussion, it need only be said that there is no macula basilaris nor any sign of a cochlea. Neither is there any rudiment of a round or oval window.

In the reptiles, it may be said that, so far as direct ancestry of the mammals goes, the labyrinth for the first time gives evidence of having acquired a new function, the sense of hearing, and the corresponding anatomical changes, therefore, are almost universally found in this class. Thus, from the macula lagena there has been evolved another neuro-epithelial area, the macula basilaris, which is destined to become the basilar membrane and organ of Corti of the more highly specialised reptiles, the birds, and the mammals.

The fenestra ovalis, opening into the tympanic cavity, is present in reptiles, with the exception of a few, such as the snake; and even in these the absence of the opening is clearly the result of retrograde changes, and, therefore, does not affect the present issue. Even in the most primitive of the living reptiles, Sphenodon, the oval window is well developed, and is closed by the footplate of the columella, as in other reptiles and in birds. I have, myself, verified this by dissection. The round window also, for the first time makes its appearance in the reptile; but although it is homologous with the same structure in mammals, it has to undergo remarkable evolutionary vagaries before it reaches the shape and situation in which it is found in the higher mammalian vertebrates.[1] It is not at present necessary to enter into details concerning the changes.

Now although the basilar membrane and organ of Corti are definitely present in reptiles, it is to be noted that these are, in most cases, so small that the bone or cartilage surrounding the whole labyrinth has had to undergo hardly any change to accommodate them. Thus, in Sphenodon and in the Chelonia there is only a slight bulging of the capsule of the labyrinth to represent what is ultimately to become the bony cochlea. In the lizards the bony capsule has been displaced a little more by the growing cochlea, and in one of the lizards, the teguixin, the bone has been tunnelled out into a definite tube to accommodate the organ. In the very highly specialised crocodilia the bony tube of the cochlea is still larger and is definitely bent on itself.

Now, while these remarkable changes are going on in respect to the oval window and the cochlea, the other parts of the labyrinth, the vestibule and the canals, remain practically stationary. And the same is true of the bony or cartilaginous capsule which surrounds these parts of the labyrinth.

It is not necessary to enter into a description of the evolution of the labyrinth in the birds, as they are not in the direct mammalian descent.

Among the mammals themselves, the monotremata offer a remarkable similarity to the reptiles. The organ in the monotremes was first studied by Pritchard, who showed that, though the organ of Corti was not of complete mammalian type, still it approached more closely to that than to the type found in the reptile. He also showed that the tube of the cochlea, though proportionately larger than in the reptiles, was only beginning to show the appearance of the spiral curvature which is so characteristic of the organ found in other mammals. In respect to the semicircular canals and vestibule these structures show no advance from the condition in which they are found in the reptile.

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[1] Gray, *Proc. Roy. Soc.*, B. Vol. 80, 1908.

Passing to the eutherian mammals, it is found that the cochlea is an organ still undergoing a progressive evolution, and in consequence causing alterations in the bone which surrounds it. The tube, increasing in length, takes on a spiral form, and the bone is channelled out into a similar shape. There is no need to enter into details further in respect to the cochlea of the mammal. There are differences in shape, size, and richness of nerve supply. In respect to the last of these, it has been shown that among the animals examined, the human cochlea is supplied by the largest number of nerve-fibres. This point, however, must not be allowed too much weight, because, in regard to this matter, the number of species examined is very small. It is important to add that, in the eutherian mammals, the organ of Corti has undergone still further specialisation, as compared with that found in the reptile and monotreme mammal.

The vestibule and semicircular canals show no further development beyond that found in the monotreme and reptile.

The oval window of the eutherian mammal differs from that of the monotreme in that it is really oval and not round, as in the latter.

Compressing into a sentence, then, the facts shortly described above, it may be said that, so far as the direct ancestry of mammals is concerned, the semicircular canals and vestibule reached their full development in the fish, and have made no further advance. The oval and round windows and the ossicles, as well as the cochlea, on the other hand, have all been evolved certainly since the first appearance of the amphibians, and very possibly may even have had no existence before the appearance of the reptiles. These latter structures, therefore, have been evolved at a period of time measured in millions of years later than the vestibule and canals.

Leaving for a moment the consideration of the evolution of the differing parts of the labyrinth, it is necessary to refer again to the relationship between otosclerosis and heredity. It has been shown, and is indeed well known to every aurist, that hereditary tendency plays a considerable part in the incidence of the disease. At the same time there is no doubt that not uncommonly no such tendency is discoverable. Nay, even in a single family tree it has been shown that otosclero is may make its appearance in an individual in whom no hereditary tendency can be discovered. Now, in previous times this might have been looked upon as an example of the transmission of an acquired characteristic. But even among those biologists who may still believe in the transmission of acquired character in one sense of the term, there is probably none who would maintain that such a condition as otosclerosis, in all its completeness and complexity, could be transmitted so faithfully in one generation as a pure modification resulting from environment.

I venture to think there is only one explanation of these facts that the biologist will be willing to accept, and that is that otosclerosis is a variation, using the term in its biological sense. Hence, the condition may arise apparently spontaneously, and yet be transmitted; so that this view of the subject enables us to understand why, although a large number of cases are obviously hereditary, yet there remain not a few in which the affection apparently arises *de novo*. Again, the well-known fact that in the inheritance of otosclerosis a generation is sometimes "skipped," now falls naturally into a category of similar examples well known to the biologist. It merely means that the tendency to variation has not actually become manifest, but has lain latent, only to be revealed in subsequent generations.

Now, in view of this explanation of otosclerosis, it is not difficult to see why the search for some single general condition of the body, as the cause of the disease, should have resulted in failure.

The primary fault, according to the explanation of the disease given in this work, is not a general one at all, but is an inherent defect in the living cells of the organ of hearing. Anæmia, pregnancy, syphilis, toxic products circulating in the blood, exposure to extreme cold, internal secretions either normal or abnormal, and all other general constitutional conditions are merely contributory causes, and there is no reason to suppose that any of them are necessary for the occurrence of otosclerosis. Similarly, local pathological changes in the middle ear, which some aurists have not hesitated to claim as being the uniform cause of otosclerosis, are not essential to the incidence of the disease.

It is doubtless true that pathological changes in the middle ear are sometimes present, as indeed has been shown in the previous pages; but it is equally certain that in a large number of cases there is no middle ear disease at all. Incidentally it should be observed that some aurists have assumed that the rosy tint over the region of the promontory, which is characteristic of some cases of otosclerosis, points to inflammatory activity in the middle ear. From post-mortem examination I have found that such is by no means the case. The rosy tint is merely due to a hyperæmia in that region, and indicates the presence of activity in the cells in the layers of the bone or cartilage below. It is in character similar to the pink coloration found at the epiphisial line of a growing bone.

The view that otosclerosis is a variation in the biological sense, and therefore innate in the individual as distinguished from an acquired modification, explains many of the curious phenomena associated with the condition.

One of the most remarkable of these is the absence in almost all cases of any interference with the function of the vestibule and semicircular canals, even in those cases in which the cochlear branch of the auditory nerve is severely affected. In the preceding pages it has been shown how the oval and round windows and the cochlea, both in regard to its bony capsule and to the nerve structures appertaining to it, are of more recent evolution than the remaining parts of the labyrinth. Now, as the biologist well knows, variations are more apt to occur in structures that are of comparatively recent origin than in those of more ancient descent. Or, to put it in other words, a structure which has remained unchanged for many ages has reached a degree of stability which is not to be expected in one of comparatively recent development, and variations are, therefore, more liable to manifest themselves in the latter. In view of these facts, therefore, the reason becomes clear why the process which we term otosclerosis selects the footplate of the stapes, the bony walls of the cochlea, and, in some cases, the nerve structures of the organ, while it almost invariably leaves the other portions of the labyrinth untouched. It is, of course, true that there are rare cases of otosclerosis in which the symptoms referable to the canals and vestibule are present. Kalenda has recorded two such cases, and in Table III, case C 2, of the present work, there is also an example of such, though of a mild type. The fact remains, however, that such symptoms are so uncommon that their absence is one of the characteristic features of the disease. And it further is a very significant fact that there is no recorded case, so far as I am aware, in which the semicircular canals or vestibule have been affected without simultaneous disturbances in the cochlea. McKenzie has also shown that the vestibular reflexes are normal in otosclerosis.

Further, a reconciliation is now possible between the views of those who, like Manasse, hold that otosclerosis is primarily an affection of the nerve-structures of the cochlea, and the view of many other aurists who look upon the condition as primarily a disease of the bone. Since the highly specialised nerve structures are evolved hand-in-hand with the bony channel which contains them, the one accommodating itself to the other, it is not at all surprising that, when a variation occurs, it should be liable to affect both structures in the same individual.

No doubt in some cases the variation manifests itself rather as a change in the nerve-structures, while in others the bony changes preponderate, but in a considerable number of cases, especially in the later registers of life, both structures are apt to be involved. There is, therefore, no real inconsistency between the views of those who hold that otosclerosis is primarily an affection of the sound-perceiving apparatus and those who look upon the changes in the bone as being the essential feature of the affection. In one case the change may be in the sound-perceiving apparatus, and in another it may be in the bony capsule, and in yet others the pathological changes may be present in both of these structures.

In the preceding pages I have employed the method of applying our knowledge of the evolution of an organ to the pathological manifestations which may occur in that organ. This is a principle which has, of course, been applied to explain certain congenital anatomical abnormalities, but so far as I am aware, it has not been employed in unravelling some of the more obscure pathological processes which are frequently encountered by the physician and surgeon. I venture to think that the method is capable of far wider application than is possible within the limits of a work which deals with the pathogenesis of otosclerosis. It might tentatively be applied in particular to those pathological problems which are not to be explained by the activity of a specific micro-organism. Thus the remarkable frequency with which carcinoma manifests itself in the breast, the rectum, and the uterus may be explained by the fact of the great changes, anatomical and physiological, which have taken place in these regions in comparatively recent periods of evolution. Another problem to which this principle might be applied, is that which deals with the occurrence of arterio-sclerosis. At present, however, these interesting pathological problems must be left for future consideration.

CONCLUSIONS

The conclusions arrived at in the foregoing pages may be summarised as follows :

1. Otosclerosis is the manifestation of a tendency to variation, using the term in its biological sense. It, therefore, possesses the qualities of variations in general. Thus, it is heritable and is frequently inherited, but at the same time it may appear apparently spontaneously, and even in such cases it is still heritable.

2. No single general constitutional condition can be

looked upon as the cause of the disease. Such conditions may, however, play an active part in making manifest a tendency which would, otherwise, have lain latent. These general constitutional conditions, which may evoke the variation, may be quite different in different cases. And it is the duty of the aurist to investigate this matter in every case with the utmost care and without bias in respect to specific conditions, such as tuberculosis, syphilis, anæmia, pregnancy, toxic absorption from the alimentary canal, or other sources of infection. In many cases no such general condition will be found.

3. The tendency to otosclerosis may be influenced by local conditions in the middle ear, Eustachian tube, or nasal passages. These conditions may be of the nature of acute or chronic inflammation. On the other hand, there is no reason whatever to suppose that middle ear, Eustachian, or nasal trouble is present in every case, or even in the majority, as some aurists have supposed.

4. In many cases the innate tendency of the cells of the organ of hearing as a whole is sufficiently pronounced that otosclerosis will become manifest without any other discoverable pathological condition, local or general. The mere physiological processes of growth and repair are sufficient to make the tendency manifest.

5. The variation may reveal itself in the nerve-structures of the cochlea or in the bony capsule of the organ, or in the footplate of the stapes and walls of the oval window. The nerve-structures of the tympanic membrane and of the external meatus are frequently affected, as is shown by the diminished sensitiveness, and by a diminution in the secretion of wax. (Whether the nerve-cells of the cerebral cortex associated with the function of hearing are also affected must at present be considered sub judice.) It may be present in one of these

situations only, or in all together. The reason of this is that these structures have all been evolved in correlation with one another and at approximately the same period of the evolution of the race.

6. The reason why the structures just mentioned are so much more liable to variation than the vestibule or the canals, is that they are of very much more recent origin and, therefore, have not reached the same degree of stability as the latter.

The bearing which these considerations have upon diagnosis, prophylaxis, and treatment is in many respects obvious, but this aspect of the subject will be discussed in another chapter.

CHAPTER IV

PATHOLOGICAL ANATOMY

One of the numerous difficulties attending the investigation of otosclerosis arises from the fact that even when the temporal bones of subjects who have suffered from the disease are obtained, they can practically never be fresh, and the cells have for that reason undergone post-mortem changes before being fixed. So far as one can foresee, this difficulty will always remain.

But even when the bones have been obtained, there is the further difficulty of their preparation for microscopic purposes. This difficulty has in large part been overcome, thanks to the investigations of Politzer, Alexander, Siebenman, Bruhl, Manasse, and others. In spite of these investigations, no satisfactory method has been described of making the microscopic sections sufficiently thin for the proper study of the finer details. The embedding medium used by all these observers was celloidin, and sections obtained by this method are at the best rather thick, though in other respects they may be admirable. For this reason the writer has devised a method of microscopic preparation more satisfactory in some respects, though less so in others. The ordinary methods of decalcification also are not very satisfactory, and the writer therefore devised a new one.

A description of these methods is, therefore, desirable in the interests of those who may intend to work upon the anatomy and pathology of the organ of hearing.

After the bones have been removed from the skull, the preparation is reduced by means of a fretsaw to the smallest possible size consistent with absence of injury to the parts to be examined. Before doing this, however, a very careful examination of the middle ear and ossicles should be made by means of a magnifying lens, and any abnormality in the way of exostoses changes in the muco-periosteum should be noted in writing. Above all it is important to observe whether the stapes appears to be fixed or moveable. It is desirable also to obtain a photograph of the promontory and inner wall of the tympanum at this stage.

A small hole is then filed in the convexity of the superior semicircular canal, another in the horizontal canal, and a third in the lower whorl of the cochlea at the point at which it is nearest to the middle line. This last puncture must be very minute, and the moment the fluid is seen coming through it from the cochlea the preparation must be put into the fixing fluid.

The fixing fluid consists of a solution of 5 parts of formalin (Schering's) with 95 parts of 80% alcohol. The amount of fluid used should be considerable, and the bone should be suspended near the surface by a thread. The time given for fixation should be not less than three days, and it is preferable to give a week. Washing out should be done in about 70% alcohol, and need not take more than a few hours.

The preparation is next put into the decalcifying fluid. This consists of fuming nitric acid 5 parts, formalin 5 parts, water 90 parts. The solution must be prepared as follows: a 10% solution of nitric acid in water is made up and kept in stock in large quantities, and similarly a 10% solution of formalin in water is kept in stock. When required, equal parts of these solutions are mixed, thus giving 5% solutions of each. The mixture must be allowed to stand for a few hours or so, and the preparation is then put in for decalcification. The reason for this delay is that if the preparation is put in immediately the solutions are mixed, the decalcification process is too rapid and it may damage the delicate parts of the specimen. I am indebted to Dr. G. J. Jenkins for this information, and I have found it to be of value.

On the other hand, as stated above, the mixed solution must not be kept in stock because decomposition occurs slowly and the decalcifying properties become much impaired. For the same reason it is important that the solution be changed frequently ; at first every twenty-four hours and afterwards twice a week at least. Another very important matter is that the bone should be suspended near the surface of the fluid by a thread. This allows diffusion to take place, and decalcification proceeds more rapidly.

The decalcification must be very complete, and usually takes two months or even more. The period varies according to the amount of fluid used and the frequency with which it is changed. After decalcification the preparation is washed out, first in running water for two or three hours and then in 70% alcohol for a week or two, the alcohol being frequently changed, and a few grains of lithium carbonate added at each change. When blue litmus paper applied to the surface of the preparation no longer turns red, the elimination of the acid may be considered sufficiently complete.

One of the difficulties very frequently encountered by the anatomist and pathologist when making microscopic preparations of the temporal bone is the occurrence of air-bubbles within the labyrinth. It might be supposed that this is a minor trouble, but in reality the presence of an air-bubble in the cochlea will spoil the organ for microscopic examination. I have frequently been consulted with regard to this difficulty, and in my earlier work I could find no way of overcoming it.

The bubbles arise from the decalcifying action of the nitric acid used in the preparation. At one time I was of opinion that it might result from evaporation of the ether in the celloidin solution used in embedding, but I have since found this view to be incorrect, certainly in the great majority of cases, if not in all; for if the preparation be put into a clearing agent, such as cedarwood oil or beechwood creosote, before the celloidin embedding takes place, the bubbles will be found to be already present. I therefore devised the following artifice, and since employing it I have had no trouble with air-bubbles.

Assuming that decalcification has been thorough, and that the acid has been completely washed out, the preparation is put into a solution of 5% formalin in water for a day or two.

A large test tube, at least one inch in diameter, is then filled with tap water and boiled for twenty minutes or half an hour. This will drive off all, or nearly all, the oxygen, nitrogen, and carbon dioxide which are present in any water which has been exposed to the air. It will also considerably diminish the amount of water in the test tube, but enough will still remain for the purpose. The bunsen flame is then turned out, and a layer of heavy paraffin oil (previously warmed by putting the bottle into warm water) is immediately poured on to the surface of the water in the test tube. ("Burroughs, Wellcome's" parolein suits admirably for this purpose.) The layer of oil should be about an inch thick. This prevents the absorption by the water of any gases from the air. The tube containing the water and parolein is then allowed to cool thoroughly. The bone is then dropped into the test tube and sinks at once to the bottom. In the course of twenty-four hours the water will have absorbed any gas that may be present within the labyrinth, and the preparation of the object may be proceeded with. The specimen obtained from Cases III and IV were prepared in this way.

The next stage is identical with that employed in celloidin embedding. The preparation is placed for forty-eight hours in 90% alcohol, then for twenty-four hours in absolute alcohol, then for twenty-four hours in a mixture of equal parts of ether and alcohol. It is then put into thin celloidin solution for two days, then into solution of medium thickness for two days, and next into thick celloidin solution for two or three weeks. The celloidin, with the preparation in it, is then hardened in the usual way, either in 85% alcohol or in the vapour of chloroform.

After hardening, the block of celloidin is pared down to the bone on all sides, except on that surface on which the stapes, the round window, the promontory, are to be found. On this surface a layer of celloidin, about one millimetre thick, is left. The object of this is, of course, to prevent injury, and to give support to the mucous lining of the promontory, the round window, the stapes, and the stapedius tendon. The paring away must be rapidly done in order to prevent the celloidin from becoming too hard as the spirit evaporates. In fact it is perhaps better to do the paring down after the next step, which consists in placing the preparation in beechwood creosote in order to get rid of the alcohol or chloroform, as the case may be.

The preparation is kept in beechwood creosote for two or three days, and the creosote is changed once. This gets rid of the chloroform or alcohol and any water which is present in the alcohol. It is then put into cedarwood oil for at least a week, the oil being changed twice. (It is very important that the cedarwood oil should be quite free of water. This may be achieved by adding anhydrous sulphate of copper to the oil.)

If the celloidin has been hardened in chloroform, the creosote stage may be omitted, the preparation being transferred direct, after paring down, to cedarwood oil.

The process of paraffin infiltration is then begun by adding small pieces of paraffin (45° C. melting-point) to the cedarwood oil in which the preparation is soaking. It is important to notice that the paraffin must not be added in such large quantities as to cause solidification, for then penetration ceases.

The preparation is kept in the cedarwood oil and paraffin wax solution for at least a fortnight, the object of this being to enable the paraffin to penetrate as well as possible at first, without the aid of heat.

The next step is to replace the cedarwood oil by chloroform, while still retaining the paraffin in the preparation. This is done by putting the latter into a saturated solution of paraffin (45° C. melting-point) in chloroform, the solution being changed once or twice in the course of two days. From this solution the preparation is transferred to the pure melted paraffin (45° C.) in the oven. The temperature of the oven must be kept accurately at 45° C. or at most 46°.

The preparation should not be kept in the melted paraffin more than four hours, and the paraffin should be changed once at least. If left much longer in the melted paraffin, the heat tends to harden and contract the celloidin, and this cannot be remedied afterwards. The tissues themselves are by this time very hard, but they, on the other hand, can be softened, as will be described later.

Embedding next takes place in the usual way with paraffin, the preparation being oriented as the paraffin hardens. It is then mounted on a "stabilit" block, care being taken to have it properly oriented. The edges of the paraffin mass are then pared down as far as possible, consistent with the edges being at right angles to one another to permit of serial section cutting.

It might be thought that the preparation would now be ready for cutting, but one more step is necessary. The decalcified bone is very hard, and it is necessary to soften it. This is done as follows. The paraffin is cut away, in a smooth horizontal plane, until the upper surface of the preparation is reached. Several layers of the preparation are then removed, leaving a flat surface of paraffin with a considerable surface of the tissues of the preparation exposed on it. The whole block is now transferred to the following solution :—alcohol (60 or 70%) 2 parts, glycerin 1 part. In the course of a few days the solution has softened the tissues on the surface and the latter stand out in relief above the paraffin.

The process of cutting the sections is now begun. The solution is wiped from the surfaces of the block which is then put in the microtome ; and sections are cut with the knife at right angles, as by the ordinary method.

It is not advisable to cut more than one millimetre in thickness of the preparation at one sitting, because the alcohol and glycerin solution does not penetrate deeper than that and the tissues become too hard for the knife. Therefore, after a series of sections has been made from the uppermost millimetre, the block is removed from the microtome and replaced in the alcohol and glycerin solution, where it is allowed to soak for a day or two. Another series of the uppermost millimetre is then cut and mounted, and the block again replaced in the solution, and so on until the whole preparation has been cut.

For staining also a new method has been devised in order to obviate the difficulty which arises when thin sections of decalcified bone are stained in watery solutions. Watery solutions cause such marked swelling of the cartilage that the sections are apt to float off the slide. The staining, therefore, is done as follows. The slides, with the sections adhering, are put for a few minutes into a bath of one-eighth per cent. solution of iron alum in 70% or [illegible]% alcohol. They are then washed out thoroughly for a minute or two in 70% alcohol. Next they are transferred to a bath of one-tenth per cent. solution of hæmatoxalin in 70% alcohol. After this they are again thoroughly washed out in 70% alcohol. From this they are dehydrated in 95% alcohol, cleared in beechwood creosote or a mixture of equal parts of beechwood creosote and xylol, and are mounted in canada balsam.

Other stains can be used, of course, for special purposes, but it must be remembered that anilin stains give rather unsatisfactory results in photo-micrography.

It will be gathered from the above that the whole process is slow and requires great care and patience. When the sections come to be examined, however, it will be found that the time and patience have been well spent, for they are superior to those prepared by any of the purely celloidin processes. In fact the method just described is the only way by which I have found it possible to get sections of the temporal bone sufficiently thin for satisfactory examination by the higher powers of the microscope. Further, the thinness of the sections permits of photo-micrography even by the higher powers of the microscope. The objections to this method are the time and patience required, and the risk of destroying the preparation during the numerous processes entailed. The older process is certainly safer, but the sections obtained are not so good.

During the past four years the writer has had the opportunity of making post-mortem examinations on four cases of otosclerosis, three of which had been examined during life. Two of the cases were clinically in a comparatively early stage of the disease at the time of death, and the significance of the pathological changes revealed by the post-mortem are, therefore, particularly interesting.

In each case the clinical report will first be given, and the anatomical changes then described.

Case I (Figs. 1–5)

Mrs. C , æt. 35. Has suffered from phthisis pulmonalis for the last two years. Dulness of hearing was first noticed three years ago, when the patient was apparently in perfect health. The onset of the deafness was gradual, and unaccompanied by any other symptom. The patient's general condition at present is comparatively good, in spite of the lung disease. There is no wasting nor rise of temperature nor night sweats.

With regard to the family history, no record of deafness could be discovered among the families of either of the parents, or among any of the patient's cousins.

At rare intervals the patient suffers from tinnitus, which is of the character of bells ringing. The tinnitus only lasts for a few minutes; otherwise there are no other subjective sound sensations. Paracusis Willisii has never been noticed.

On examination the membranes are almost normal in appearance, but there is perhaps a slight indrawing. There is no pink tinge over the region of the promontory.

The whispered voice (normally heard at 6 yards) is heard at a distance of 2 feet from the right ear, and at from 6 to 9 inches from the left ear.

Weber: Fork heard equally well in both ears.

Rinne's test: Right, − 12. Left, − 15.

Schwabach's test: Right, + 5. Left, + 6.

Low notes: Lost below ut_1 in both ears by air conduction, but all are well heard by bone conduction.

Galton's whistle: The high notes are heard up to normal range in the right ear, but in the left ear there is a very slight loss at the upper end of the scale.

Gellé's test: Bone conduction is unaffected by air pressure in either meatus.

On inflation, air passes freely into the tympanum and there is apparently a very slight improvement afterwards. This improvement is very fleeting.

Three months after the clinical examination had been made, the patient died very suddenly from hæmoptysis. The general health had continued remarkably good until this seizure, and the hearing had undergone no noticeable change.

Post-mortem Examination.—At the necropsy the bones of the head presented no abnormal appearance. Both temporal bones were removed and fixed for four days in a mixture of formalin 3 parts, rectified spirits 97 parts.

On naked-eye examination of the right ear the following conditions were found. The outer ear was normal in every respect. The middle ear also presented no abnormality, but it is interesting to note that there was a dehiscence on the floor of the tympanum, and the bulb of the jugular vein projected into the tympanic cavity. The mucous membrane was perfectly normal, and the two outer ossicles were moveable. The stapes appeared to be fixed or at least partially so, but on account of the importance of not damaging the preparation, very little force was used in testing the mobility of this ossicle. No adhesions were found anywhere in the tympanum, though such were very carefully looked for.

The left ear showed no abnormality so far as the outer ear was concerned. The middle ear contained a large amount of exuded bloody serum, obviously a condition of very recent occurrence and probably during the death-agony. On removal of this serum the mucous membrane appeared to be normal, but it was difficult to be sure of this on account of the staining effects of the bloody serum. The ossicles were normal in appearance, and the two outer ossicles were moveable. The stapes appeared to be fixed, but this point was doubtful, very little force being used in testing the degree of mobility. No adhesions were found in the tympanic cavity. There was no dehiscence of the floor of the tympanum on this, the left side.

Microscopic Examination, Right Temporal Bone.—From the series of sections cut by the method previously described, the following conditions were found.

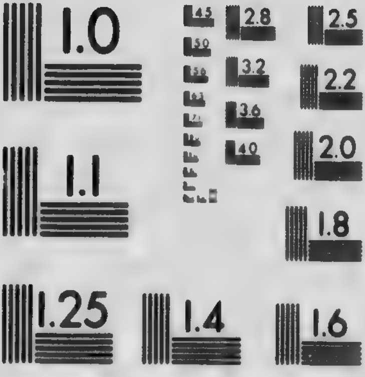
The labyrinth and the surrounding temporal bone present a perfectly normal appearance in all parts, except over a very small region in front of, above, and below the oval window. In this small affected region the bony capsule of the labyrinth has undergone that profound change characteristic of otosclerosis.

The affected region is sharply delimited from the surrounding bone and stains more deeply with hæmatoxylin. With regard to this characteristic, however, it will be seen that the staining is not equally strong throughout. Thus, as shown in fig. 2, the outer and larger portion, which lies towards the tympanic and articular surfaces, is more deeply stained than the inner and smaller portion which lies in the cleft between the vestibule and the first whorl of the cochlea. The passage from the one portion to the other is, however, gradual, being in marked contrast to the sharp line of demarcation separating the whole affected area from the normal labyrinthine capsule which has been referred to above.

It will be observed that the affected portion of bone does not quite reach the wall of the cochlea itself, and it may be added that in none of the whole series of sections does this occur. Thus, in this case there is always a layer of the normal bony capsule between the cochlea and the diseased bone, a condition which does not occur in many cases. It should also be observed that in that region where the affected bone reaches to the muco-periosteum of the middle ear, this muco-periosteum is slightly thickened. In the other portions it is of normal thickness. The thickening of the muco-periosteum shows no sign of round-celled infiltration or of inflammatory activity.

The porous character of the affected region, especially in the deeper stained portion, is in marked contrast with the dense bone of the normal capsule. The larger spaces of the affected portion have a network appearance which is due to the presence of marrow. In many of the spaces blood-vessels are seen, and it may be noted that the spaces communicate freely with one another.

On examination with the higher powers of the microscope it is found that the large spaces in the most deeply stained region of the affected area are surrounded by lamellæ, similar to those found in normal bone; a condition which is evidently due to the activity of the osteoblasts. The lamellation, however, is not perhaps quite so perfect as is found in normal bone, but the most noticeable feature is in regard to the osteoblasts themselves, which are rounder and plumper than in normal conditions. At the same time this portion presents structural appearances rather like normal spongy bone, except for the fact that it stains deeply, and that osteo-



MICROCOPY RESOLUTION TEST CHART
NATIONAL BUREAU OF STANDARDS
STANDARD REFERENCE MATERIAL 1010a
(ANSI and ISO TEST CHART No 2)

clasts are entirely, or almost entirely, absent. In this particular case it is the deeply stained portion which is nearest to the oval window and, indeed, comes actually up to the wall of the latter.

On passing from this more deeply stained portion to that which is less deeply stained, a gradual change is noticed in several respects. Besides the fact that the latter portion stains less deeply, it is found that the large spaces become fewer in number. Further, the tissue between the spaces is much less rich in small cells and more homogeneous in character, and osteoclasts are present. The most striking features, however, are the absence of cells which can be called osteoblasts, and the absence of any lamellation. Near the sharp line of demarcation which separates the less deeply stained portion of the diseased area from the still more faintly staining normal bony capsule the number of large cells (osteoclasts) is increased. These cells of the osteoclast type frequently lie in actual contact with the normal bone of the capsule. The present writer's findings in this respect do not agree with those of Manasse in general, though that authority admits that perhaps in some cases osteoclasts may be found absorbing the bony tissue of the capsule just outside the diseased area.

It is interesting to observe that there is no sign of any inflammatory process anywhere, either in the area of new-formed bone or in the neighbouring portion of the normal bony capsule. Even the slight thickening of the muco-periosteum over the diseased bone near the oval window shows no evidence of round-celled infiltration or other signs of inflammatory activity.

Before leaving the description of the change in the bony capsule, it is to be noted that the area of new-formed spongy bone extends right up to the stapedio-vestibular articulation, but at no point in any of the sections does it pass beyond this limit. Thus the stapes remains free from bony union. At the same time it may be seen in some of the sections that the new-formed bone has expanded both internally and externally to the tip of the footplate of the stapes, so that the latter lies in a groove in the oval window. Hence the effect of this change would be to impede the movements of the stapes to a certain extent. This effect would be increased by the thickening of the muco-periosteum of the tympanum in the immediate neighbourhood of the footplate of the stapes.

The tensor tympani and stapedius muscles are quite normal.

Membranous Structures.—The soft structures of the labyrinth do not show any sign of pathological change. There is, of course, as is inevitable, much evidence of post-mortem change, but there is none which can be looked upon as even suggestive of a diseased condition. Thus the organ of Corti differs from the normal structure as seen in freshly fixed specimens (such as can only be obtained from animals) only in regard to the obviously post-mortem disintegration of the hair cells, supporting cells and cells of Hensen. Although the hair cells have undergone disintegration, the hairs themselves remain as under normal conditions. The tunnel of Corti is quite normally arched, there are no adhesions either of the tectorial membrane or of Reissner's membrane to the upper surface of the organ. The ligamentum spirale shows no sign of atrophy or degeneration, and the stria vascularis is perfectly healthy in appearance. The ganglion cells of the ganglion spirale show no departure from the normal beyond those characteristic of post-mortem changes. Finally, the cochlear and other portions of the auditory nerve appear to be quite healthy.

All the soft structures, including the nerves of the internal ear, present a normal appearance.

Microscopic Appearance in the Left Ear.—The appearance presented by the left ear reveals the extraordinary symmetry, both in extent and character, of the disease. In fact a description of the appearances on the left side is little more than a recapitulation of those already described on the right side.

The affected area, which measures about 1 mm. in diameter, is sharply demarcated from the surrounding normal bony capsule. Posteriorly it reaches right up to, but does not actually pass, the margin of the oval window. There are no cartilage cells remaining on the vestibular aspect of the stapedio-vestibular articulation. On its outer aspect the diseased area extends up to, and lies in contact with the muco-periosteum of the tympanic cavity, and in this region the muco-periosteum is slightly thickened. In other portions this membrane is normal. Anteriorly and internally the diseased focus reaches as far as the membranous wall of the cochlea, so that over a very small area the two are in contact. In this respect, therefore, the left side differs from the right.

With regard to the details of the changes in the diseased focus, these are very similar to those found on the right side.

There is a posterior and external portion which stains particularly deeply, and an anterior and internal portion which stains rather less deeply than the former, but more deeply than the adjacent normal bony tissue of the capsule. Of these two portions, the outer, more porous and more deeply staining, is much larger than the inner and less deeply staining portion. The more deeply staining portion shows comparatively good formation of new bone; that is to say, the lamellation is regular and similar to that found in normal bone. The osteoblasts, however,

Case I—Fig. 1

RIGHT EAR

Horizontal Section through the Petrous Portion of the Temporal Bone. × 8

f. Facial nerve.
h. Horizontal semicircular canal.
p. Posterior semicircular canal.
m. Internal auditory meatus.
v. Vestibule.
f. Footplate of stapes.
s. Stapedio-vestibular articulation. There is no ankylosis.
n. Focus of diseased porous bone in front of the oval window.

[illegible]

[illegible]

Horizontal Section through the Petrous Portion of the Temporal Bone. [illegible]

f. Facial nerve.
h. Horizontal semicircular canal.
p. Posterior semicircular canal.
m. Internal auditory meatus.
v. Vestibule.
j. Footplate of stapes.
s. [illegible]
e. [illegible]

m
h
f
g
n
d
e
j

Case I—Fig. 2

RIGHT EAR

Section through the Focus of Diseased Porous Bone. × 80 ca.

o, *o*. Old normal bone of the capsule of the labyrinth.

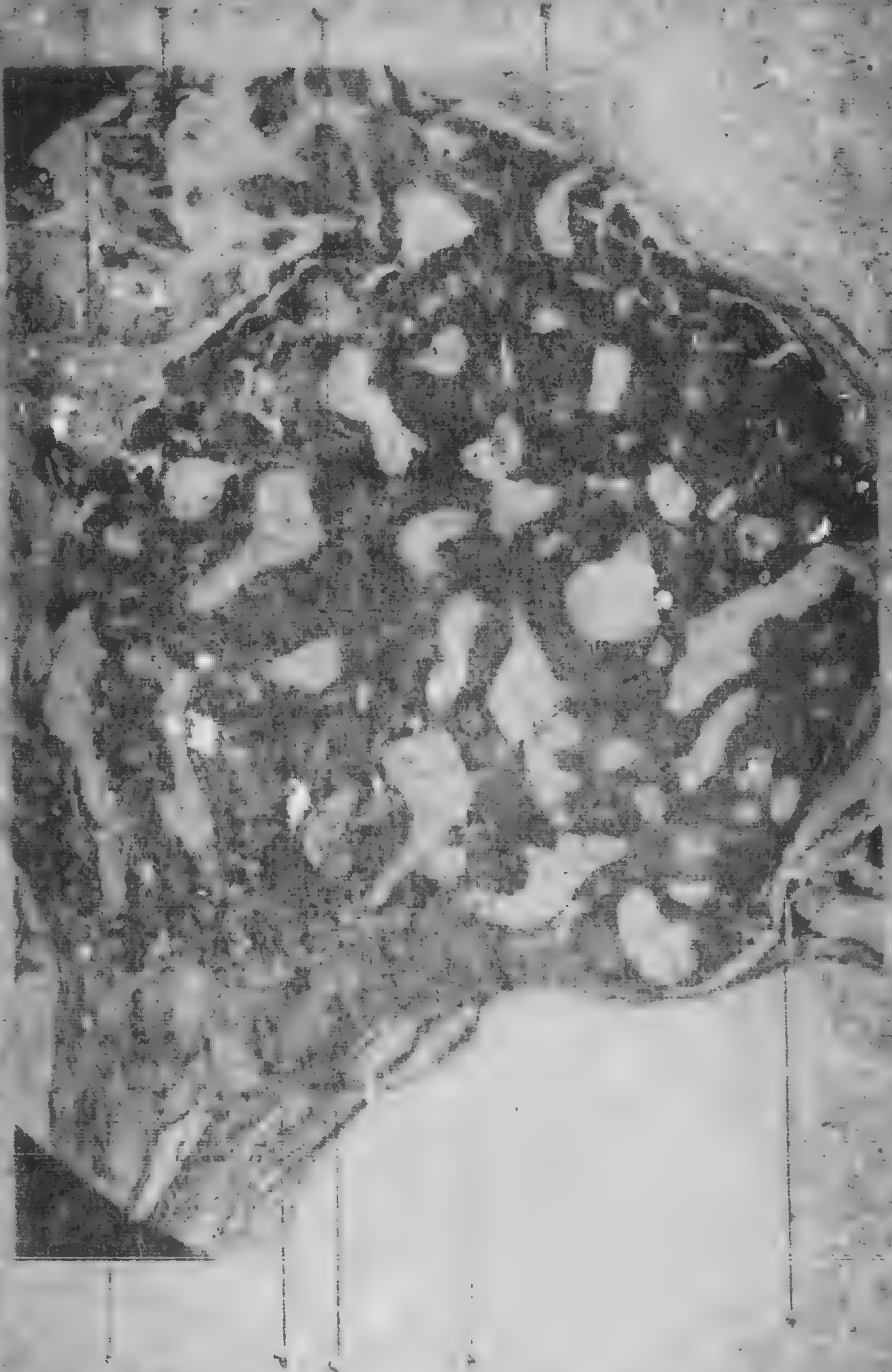
d, *d*. Line of demarcation.

f, *f*. Focus of diseased porous bone. It is highly vascular, and the spaces contain marrow in considerable quantities. The top left-hand portion of the photograph shows the portion of diseased bone which is stained rather more faintly than the rest. The same peculiarity is found in the left ear (see fig. 5). The diseased bone is laminated like ordinary bone.

v. Vestibule.

s. Stapedio-vestibular articulation. There is no ankylosis, but the diseased bone reaches to the articular surface of the oval window.

m. Mucous lining of tympanum.



[illegible]

[illegible]

Necrosis [illegible]

c, c. **Old** [illegible]

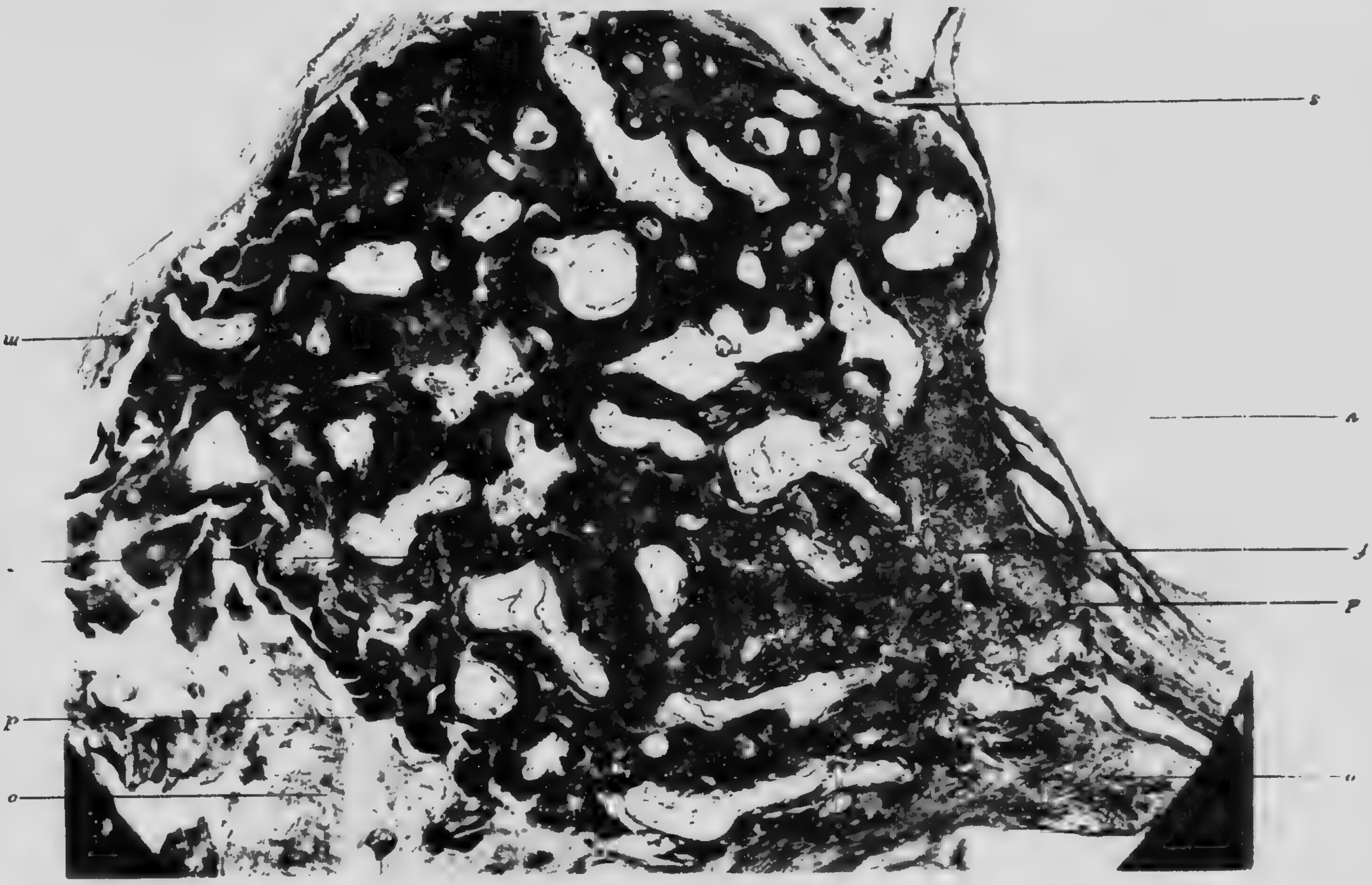
d, d. Line of demarcation.

f, f. **Focus** of diseased porous bone, [illegible] vascular, [illegible] spaces [illegible] marrow [illegible]. The top left-hand portion of the photograph [illegible] portion of diseased bone [illegible] the rest. [illegible] The diseased [illegible]

[illegible] **Vessels** [illegible]

[illegible] **Shows** [illegible]

[illegible] **M**[illegible]


w
p
o
s
a
f
p

Case I—Fig. 3

RIGHT EAR

Section across the Line of Demarcation and Adjacent Regions

× 170 ca.

To the left and below is the faintly-stained old normal bone of the labyrinthine capsule. In the upper part of the photograph is shown a portion of the diseased focus of bone. This diseased portion is divided into two regions, right and left. The region to the right does not stain deeply, and the cellular elements of which it is composed are not very sharply defined. It consists of irregular-shaped protoplasmic masses with many nuclei; these are giant-cells or osteoclasts. They are formed, according to Kölliker, by the fusion of the osteoblasts. They have absorbed the mineral and other components of the bone, and consequently lamellation is quite absent. This is the most recently affected portion of the bone, and in it there is no deposit of new bone nor are marrow-spaces present. The darker stained region above and to the left of that just described shows the stage in which new bone is beginning to be deposited and marrow-spaces are being formed. Osteoblasts are seen, but the new-formed bone does not yet possess the lamellated structure of normal bone. The latter may be seen in fig. 2.

c, c. Normal bone of capsule of the labyrinth.
d, d. Line of demarcation.
a. Region in which the bone of the capsule has been absorbed, but in which, as yet, no new bone has been deposited.
b. Region in which new bone is being deposited, hence the deeper staining. There is, however, no lamellation.
g, g Giant-cells or osteoclasts in the region in which the old bone has been absorbed.

[illegible]

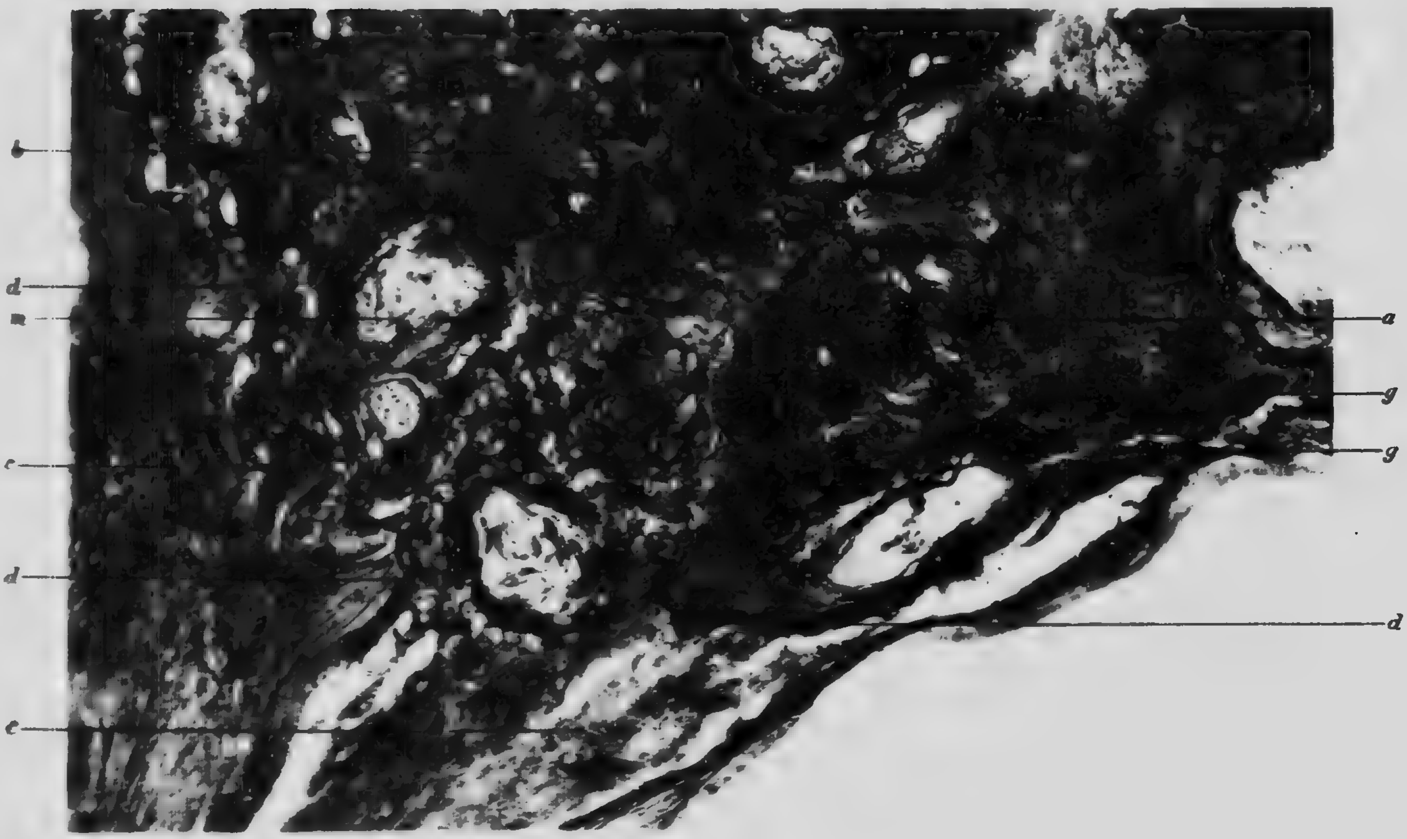
[illegible]

SECTION ACROSS [illegible]

To the left and [illegible] the [illegible] intermediate [illegible] shows a [illegible] as [illegible] that [illegible] period [illegible] protoplasm [illegible] clear [illegible] the [illegible] ments of the [illegible] This is the most [illegible] is no [illegible] **stained** [illegible] stage in which [illegible] spaces are [illegible]

[illegible]

1. [illegible]
2. [illegible]
   - a [illegible]
   - b [illegible]
   - c [illegible]

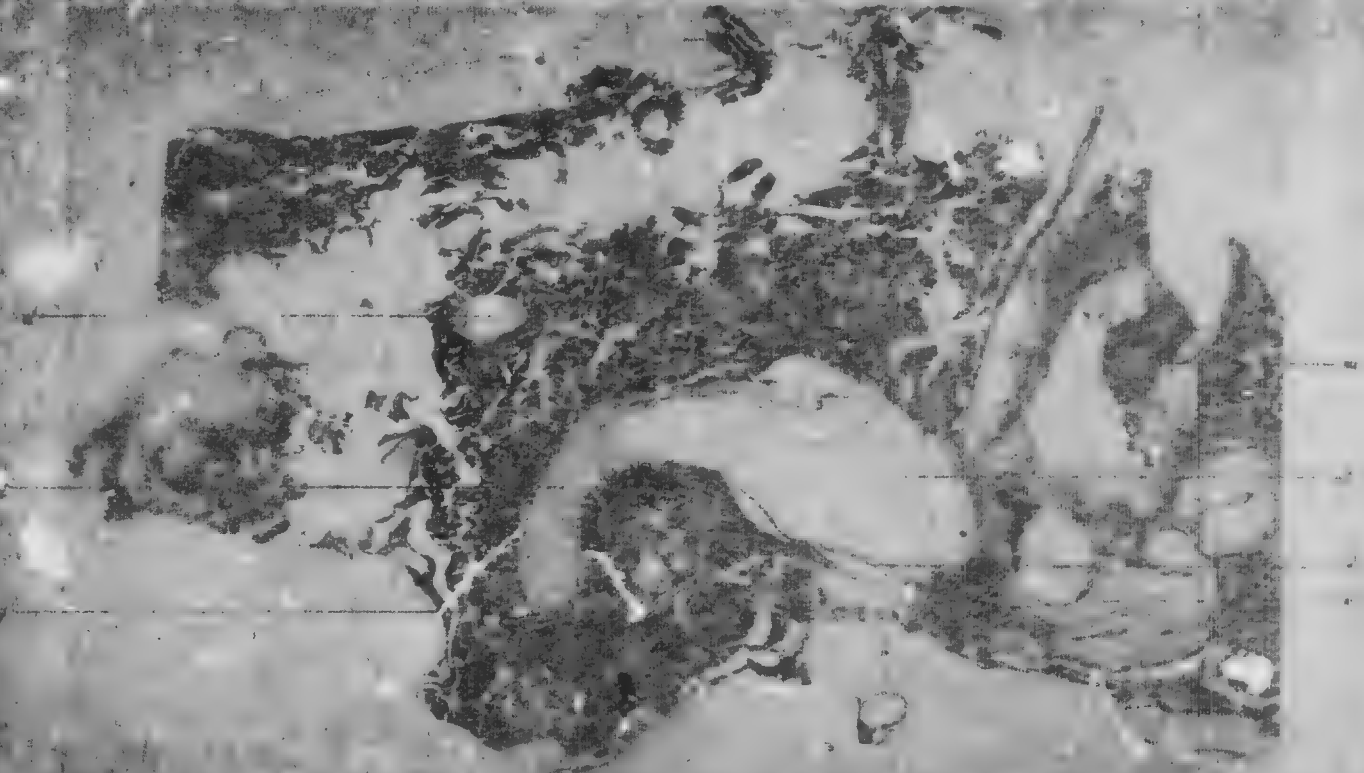

b
d
m
c
d
e
a
g
g
d

Case I—Fig. 4

LEFT EAR

Horizontal Section through the Petrous Portion of the Temporal Bone. × 8

n. Focus of diseased porous bone. Deeply stained.
j. Footplate of stapes.
v. Vestibule.
m. Cochlear branch of auditory nerve, lying in the internal auditory meatus.
p. Posterior semicircular canal.
h. Horizontal semicircular canal.
f. Facial nerve.



CASE I.—FIG. 4

LEFT EAR

HORIZONTAL SE[illegible] PETROUS PORTION OF THE TEMPORAL BONE. × 5

- a. Focus of diseased porous bone. Deeply stained
- f. Footplate of stapes.
- v. Vestibule.
- n. Cochlear branch of auditory nerve, lying in the internal auditory meatus.
- p. Posterior semicircular canal
- h. Horizontal [illegible]
- f. Facial [illegible]

d
y
f

CASE I—FIG. 5

LEFT EAR

SECTION THROUGH THE FOCUS OF DISEASED POROUS BONE. THE DISEASED BONE IS STAINED DARK; THE OLD NORMAL BONE IS MORE FAINTLY STAINED. × 80 ca.

s. Stapedio-vestibular articulation. The anterior termination of the footplate of the stapes is seen. There is no bony ankylosis, but the diseased porous bone reaches to the articular surface of the oval window.

v. Vestibule.

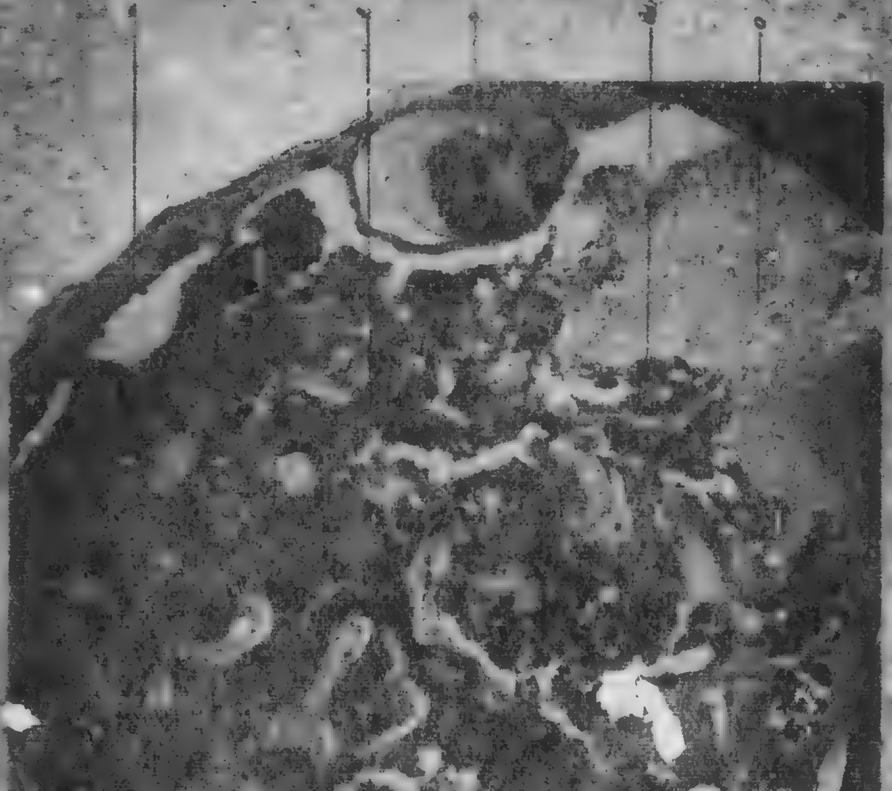
f, f. Focus of diseased porous bone. It is rich in blood-vessels, and many of the spaces contain marrow. A portion of the focus in the lower left-hand region is stained slightly paler than the rest.

d, d. Line of demarcation.

o, o. Old normal bone of the capsule of the labyrinth

t. Tympanic plexus.

m. Mucous lining of tympanum.



[illegible]—Fig. 5

[illegible]

SECTION THROUGH THE FOCUS OF [illegible]. THE DISEASED BONE IS STAINED DARK, [illegible] BONE IS MORE FAINTLY STAINED. × 80 ca.

a. Stapedio-vestibular articulation. The anterior termination of the footplate of the stapes is seen. There is no bony ankylosis, but the diseased porous bone reaches to the articular surface of the oval window.

v. Vestibule.

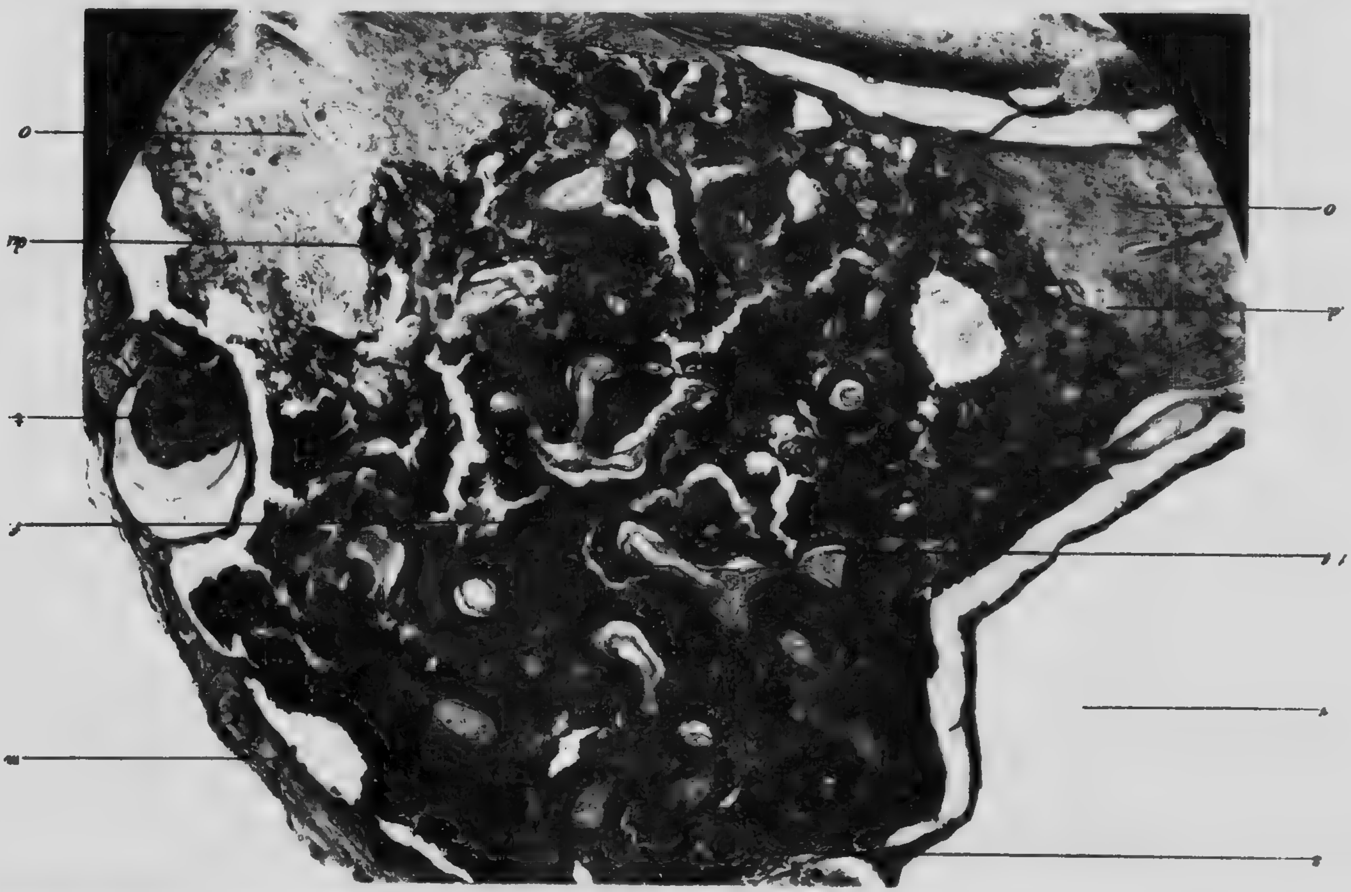
f, f. Focus of diseased porous bone. It is rich in blood-vessels and many of the spaces contain [illegible]. A portion of the focus in the [illegible] than the rest.

d, d. Line of demarcation.

o, o. Old normal bone of the capsule of the labyrinth.

t. Tympanic plexus.

m. Mucous lining of tympanum.


o
rp
t
f
m
o
p'
ti
a
s

are rounder than is normally the case in bone and the canaliculi are not so well developed. Marrow is present in the larger spaces and blood-vessels are also found in them.

In the more faintly stained portion of the diseased area the structure presents a less organised appearance. There is a considerable amount of apparently granular débris, and amongst this there are many large cells, very similar to, if not identical with, osteoclasts. There is no lamellation in this portion of the diseased area. Osteoblasts are not found.

At certain points in the line of demarcation evidently newly formed, or rather newly forming, blood-vessels may be seen passing outwards from the diseased area through the line of demarcation into the surrounding normal bony capsule.

The tensor tympani and stapedius muscles are quite normal.

In respect to the soft structures of the inner ear, there is nothing more to be said than that these present a normal appearance throughout. The organ of Corti shows signs of post-mortem disintegration exactly similar to those found in examination of normal human temporal bones which have been put in fixing fluid a considerable time after death. The changes are similar to those found on the right side and described in the preceding pages.

It must, however, be emphasised that although no pathological changes are discoverable in the nerve structures of the inner ear, this does not mean that pathological changes are necessarily absent. It only signifies that if any such changes are there, they are not revealed by our present methods of microscopical investigation.

CASE II (FIGS. 6–8)

Mrs. N., æt. 49. The patient has been the victim of phthisis pulmonalis for the last three years, and her general health has been poor. Dulness of hearing began about one year ago, with a gradual onset and without any pain or discharge. Tinnitus was first noticed a little time after the onset of the deafness, and has continued constantly ever since. It is pulsating in character, " like the throbbing of an engine." It is present in both ears, but worse in the left, and in neither ear is it distressing. Paracusis is noticeably present. Of a family of three, the patient is the only one who is dull of hearing. The father was slightly deaf, but this was attributed to boiler making. The mother was not dull of hearing, nor is the patient aware of any deafness among her uncles, aunts, or cousins.

The hearing becomes worse in wet weather. There have been one or two very slight attacks of giddiness, but this has been attributed, and probably correctly, to general ill-health, the result of the lung disease.

Examination.—A considerable amount of wax was present in both ears. After this had been removed the following conditions were found :

Right ear.—The tympanic membrane appears to be normal in every respect. There is no rosy tint present over the region of the promontory. Watch heard at a distance of 2 inches (normal distance 2½ yards). Whisper, 1½ feet (normal 6–7 yards). Conversation voice, 2½–3 yards.

Schwabach's test, + 4.

Rinne's test, − 7.

High notes, no appreciable loss.

On inflation, air enters the tympanum freely, but there is no very noticeable improvement in the hearing as a result.

Left ear.—There is a large perforation in the posterior, inferior quadrant, but the malleus is still present. There is no pus or moisture in the meatus or middle ear, but flakes of epithelial débris are seen behind the upper posterior margin of the perforation, and these prevent any view of the deeper parts in that region.

On inflation, air enters the tympanum freely, but there is no improvement thereafter.

Owing to a mistake the hearing-power of the left ear was not ascertained.

The patient died three months and five days after the examination just reported; and the post-mortem took place about twenty-four hours after death. The changes found were as follows:

Macroscopic Examination.—*Right ear.*—The outer ear is quite normal and the tympanic membrane, beyond perhaps a slightly increased translucency, is healthy in appearance. The middle ear appears to be healthy in all its parts. The mucous membrane shows no sign of thickening or of congestion, and there is no secretion in the tympanic cavity. The Eustachian tube is patent and the mucous membrane healthy. The malleus and incus are normally moveable, and there are no adhesions in the region of the oval window nor anywhere else. The stapes permits of movement in a vertical direction, but it is doubtful if there is an inward or outward movement. This point, however, is uncertain as only the slightest pressure is employed.

Left ear.—The outer ear is normal and there is no discharge in the meatus. A large perforation is seen in the posterior inferior quadrant of the tympanic membrane. On opening the tympanic cavity, it is found that all the upper part of the cavity proper is filled by cholesteatomatous débris, and this extends into the attic and antrum, filling both these cavities.

On careful removal of the cholesteatomatous mass, the malleus and incus are found to be present, and there are no adhesions limiting the movements of the ossicles. The crura of the stapes are moveable to a very slight extent in a vertical direction, but whether the ossicle is moveable horizontally in the oval window is doubtful. There are no adhesions in the neighbourhood of the stapes and oval window, nor indeed anywhere else in the tympanum. The mucous membrane is pearly grey in colour and glazed in lustre, as indeed is usually the case when cholesteatoma is present. There is no pus or moist secretion in the tympanum. The Eustachian tube is healthy and patent.

Microscopic Examination.—*Right ear.*—The microscopic examination in this case of Etosclerosis reveals a condition not hitherto described.

As in the previous case, the affected area of bone is extremely small. It is, moreover, found in the usual position, in front of, above and, to a less extent, below the oval window. This is the only focus of disease in the bone. At its widest the area measures about one millimetre.

The affected portion of bone reaches up to, but does not involve the stapedio-vestibular articulation. In front and internally it extends towards, but does not reach at any point the lowest whorl of the cochlea. Internally the diseased area forms the bony wall of a portion of the vestibule, and externally on the tympanic aspect it lies immediately under the muco-periosteum of the tympanum. At no point is the stapes fixed to the oval window by bony tissue.

It is interesting to note that the walls of the pelvis of the fenestra ovalis are not encroached upon ; that is to say, the contour of the parts is similar to that found under normal conditions. The reason for this apparently is that although the bone in this region has undergone obviously profound changes, yet there is no evidence of the occurrence of new-formed bone, such as was present in Case I, and which had encroached upon the walls of the pelvis of the oval window.

The deposition of new-formed bone in the affected area has hitherto been considered an invariable feature of otosclerosis, and the outstanding character of the change in the bone. The present case, therefore, is of special interest inasmuch as it definitely proves that such a view is incorrect.

As in all other cases of otosclerosis, the affected area is separated from the surrounding normal bone by a sharp line of demarcation. But it will be noticed that the affected area of bone stains *less* deeply than the normal bony tissue. (It may here be remarked that the whole process of preparation and staining was exactly the same as that employed in Case I.) In this respect, therefore, as in others to be described below, this case stands out in marked contrast to the first case, and indeed to all other cases of otosclerosis which have hitherto been published, so far as the writer is aware.

Under low magnification it may be noted that the blood-vessels within the affected area are large. Furthermore, although the diseased bone in the present case may be described as being more porous in texture than the surrounding normal bone, yet the porosity is very different in character from that found in Case I and in the ordinary cases of the disease hitherto described. The difference consists in this, that in the present instance the spaces in the bone are almost completely filled up by the blood-vessel whereas in Case I these spaces are only occupied by the blood-vessels to a very small extent, a greater part being filled by marrow and cellular structures.

On examination with the higher power of the microscope, somewhat different conditions may be seen in different portions of the diseased area. In the portion near the centre, the tissue consists almost entirely of a network of blood-vessels. The bony structure has almost completely disappeared by absorption, and even the cellular elements are altogether scanty, so that empty spaces of considerable size are to be seen.

In the portion near the stapedio-vestibular articulation the blood-vessels are large and numerous, but the spaces between the blood-vessels are occupied by osteoblasts and fine granular débris. A few large irregular-shaped cloudy cells are also seen in this region. The bone has to a large extent lost its lamellated appearance.

In the region near the line of demarcation the appearances are very similar to those seen near the stapedio-vestibular articulation. That is to say, the blood-vessels are large and there are numerous osteoblasts occupying the spaces between the vessels. There are also occasional large irregular-shaped cloudy cells, and there is but little lamellation. On the other hand, the granular débris which is seen in the area near the stapedio-vestibular articulation is not found in that portion near the line of demarcation.

Membranous Structures.—The soft structures of the cochlea present appearances in no way different from the normal. Of course, as is always the case in dealing with the human subject, changes are found; but these are the result of post-mortem disintegration. There is one little exception to the above statement, and that is that the blood-vessels of the modiolus are dilated to an extent greater than I have ever seen in subjects who possessed normal hearing in life.

There is no depression or bulging of the membrane of Reissner, nor is the latter adherent to the tectorial mem-

7

CASE II—FIG. 6

RIGHT EAR

HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE. × 8

n. Facial nerve.
m. Stapedius muscle.
p. Posterior semicircular canals.
b. Focus of diseased bone in front of the oval window. It stains *less* deeply than the adjacent unaffected bone, and in one part of it the process of absorption has been so marked that there is no bone left.
v. Vestibule.
a. Internal auditory meatus.
t. Tensor tympani muscle.

[illegible]

[illegible]

[illegible]

a. [illegible]

m. [illegible]

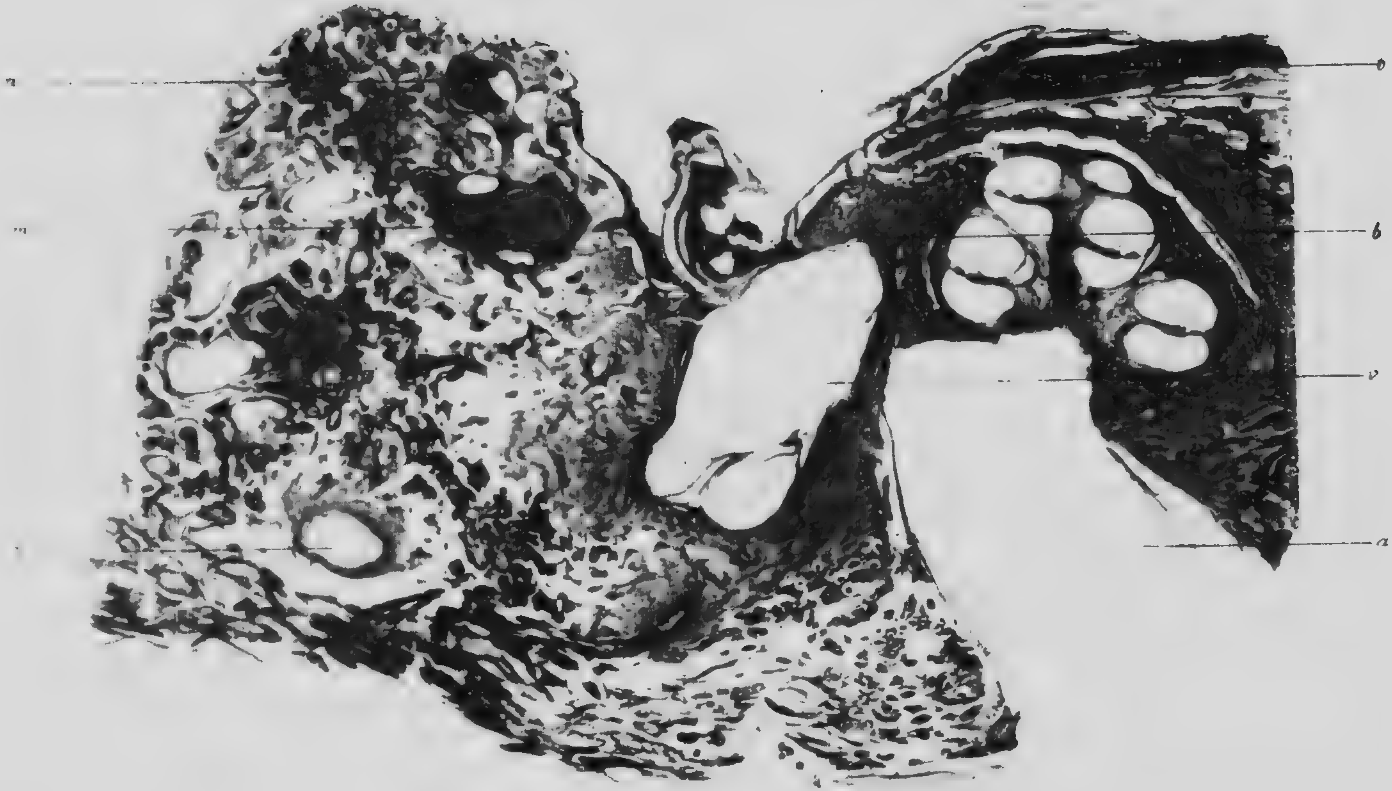
p. [illegible]

h. [illegible]

s. [illegible]

[illegible]

[illegible]


n
m
o
b
v
a

Case II—Fig. 7

RIGHT EAR

Section through the Focus of Diseased Bone. × 80 ca.

m. Mucous lining of the tympanum.

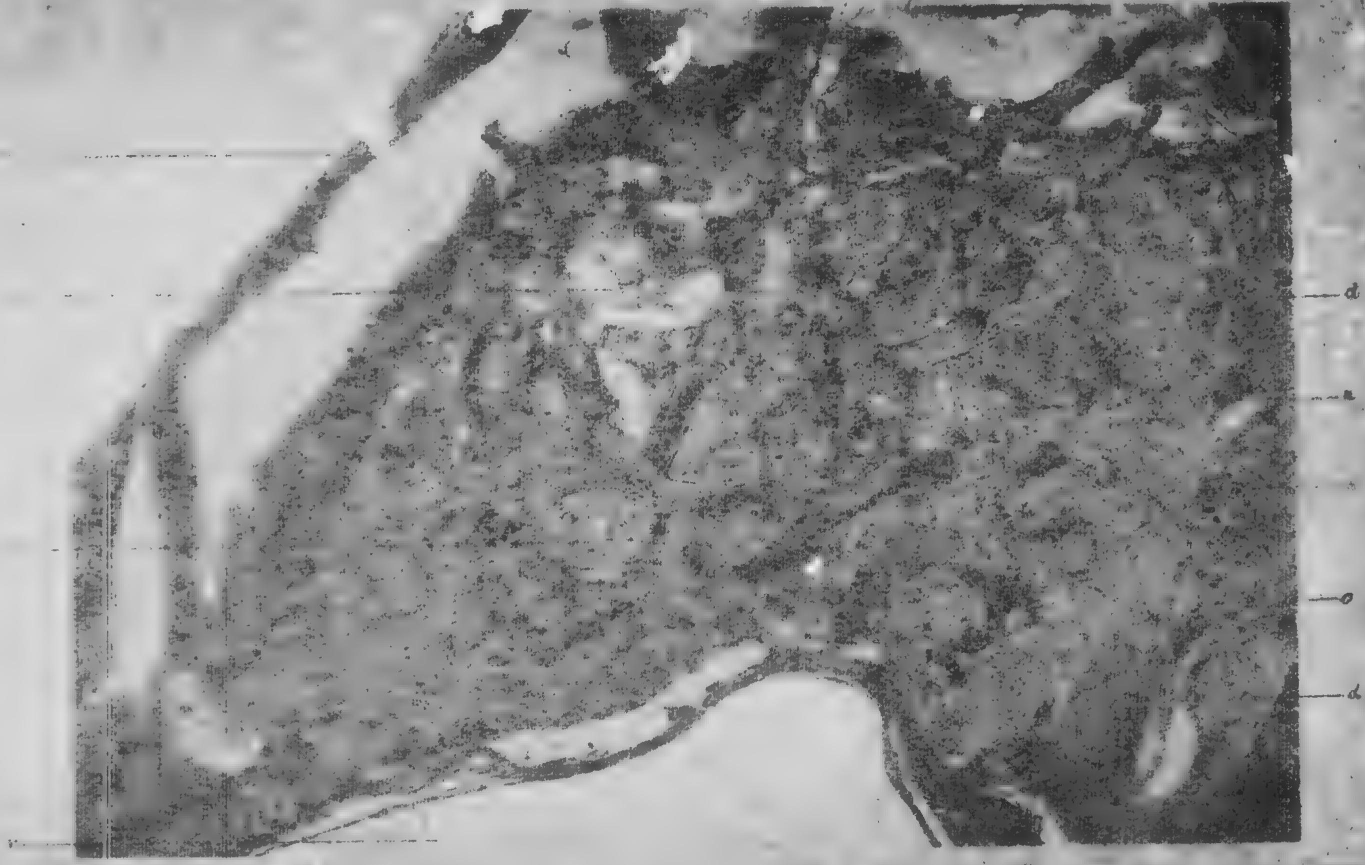
b. The pointer ends in a portion of the diseased area in which practically no bony tissue is present. Indeed, in this region there appears to be little else than a network of blood-vessels. The rest of the focus, though very vascular, consists for the most part of bony tissue in process of absorption.

n, n. Focus of diseased bone. The bone presents a laminated appearance in many parts, but the lamination is not so perfect as in normal bone.

It is of importance to note that in this case alone, out of the four examined, does the area of diseased bone stains more faintly than the normal bone of the capsule of the labyrinth.

d, d, d. Line of demarcation.

o. Old normal bone of the capsule of the labyrinth.


d
a
o
d

Case 12. P.

RIGHT EAR

SECTION [illegible] of Diseased bone, [illegible]

a. Mucous lining of the tympanum.

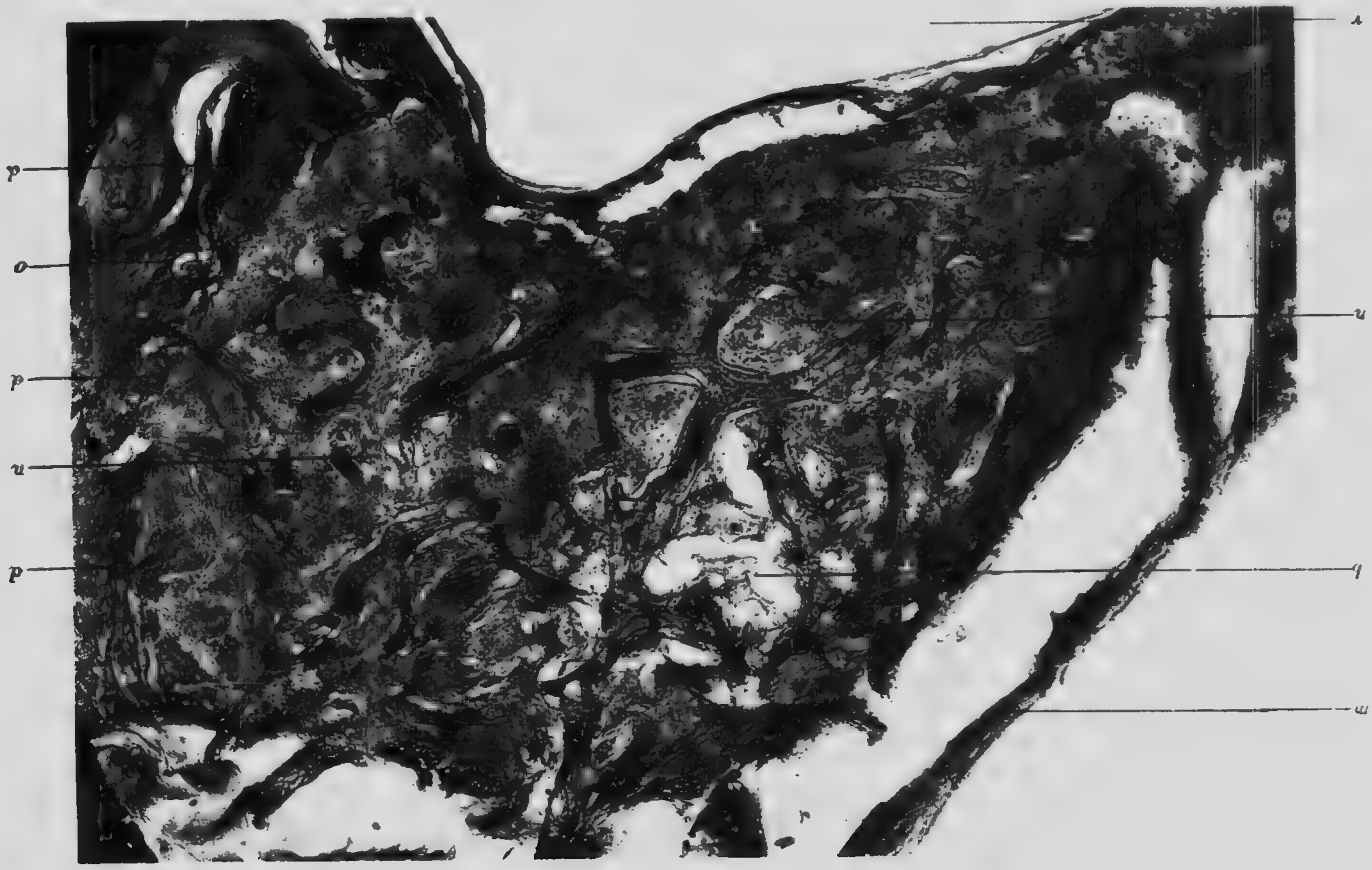
b. The [illegible] in a portion of the diseased bone, in which [illegible] [illegible] [illegible]

n, n. [illegible] [illegible]

[illegible] [illegible]

d, d, d. [illegible]

e. Old [illegible]


p
o
p
u
p
r
u
l
m

CASE II—FIG. 8

LEFT EAR

HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE. × 8

n. Focus of diseased bone in front of the oval window. It will be observed that in this case the diseased bone stains *less* deeply than the normal bone of the capsule.

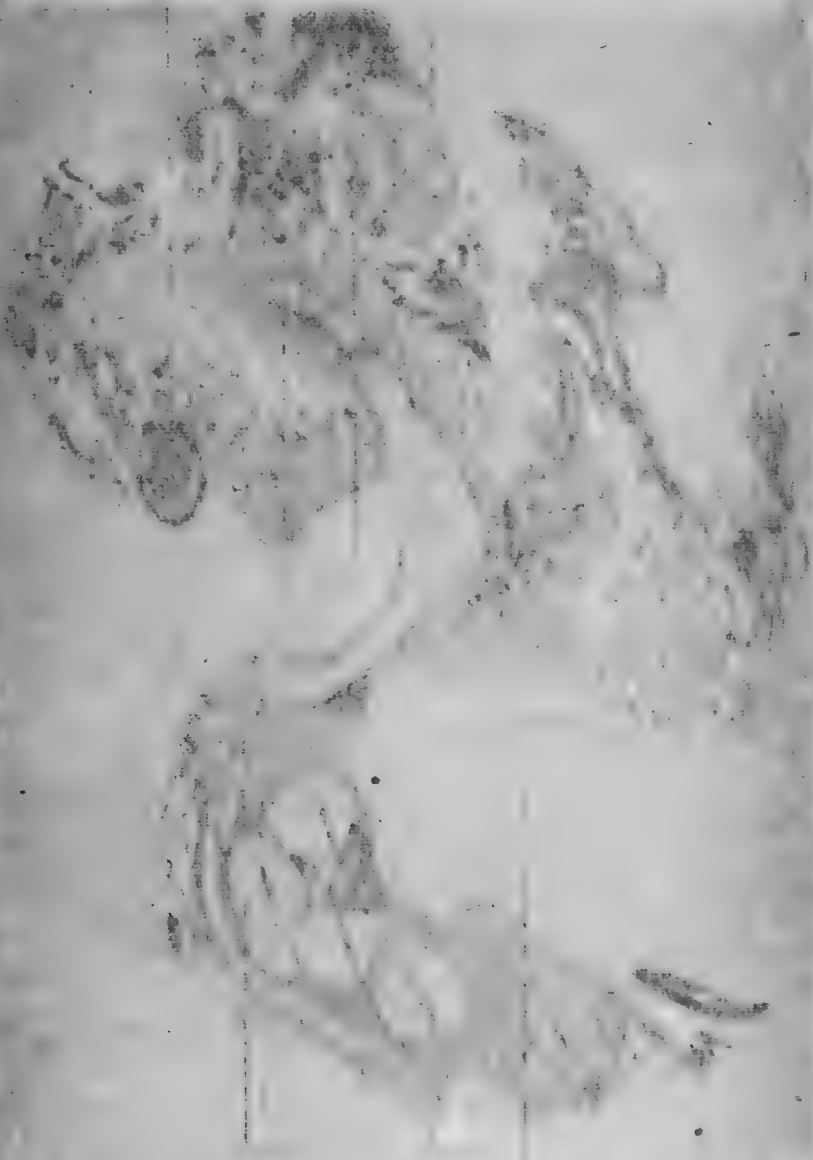
a. Internal auditory meatus.

p. Posterior semicircular canal.

v. Vestibule.

f. Footplate of stapes.

t. Facial nerve.



[illegible]

[illegible]

HORIZONTA[illegible]

[illegible]

[illegible] than [illegible]

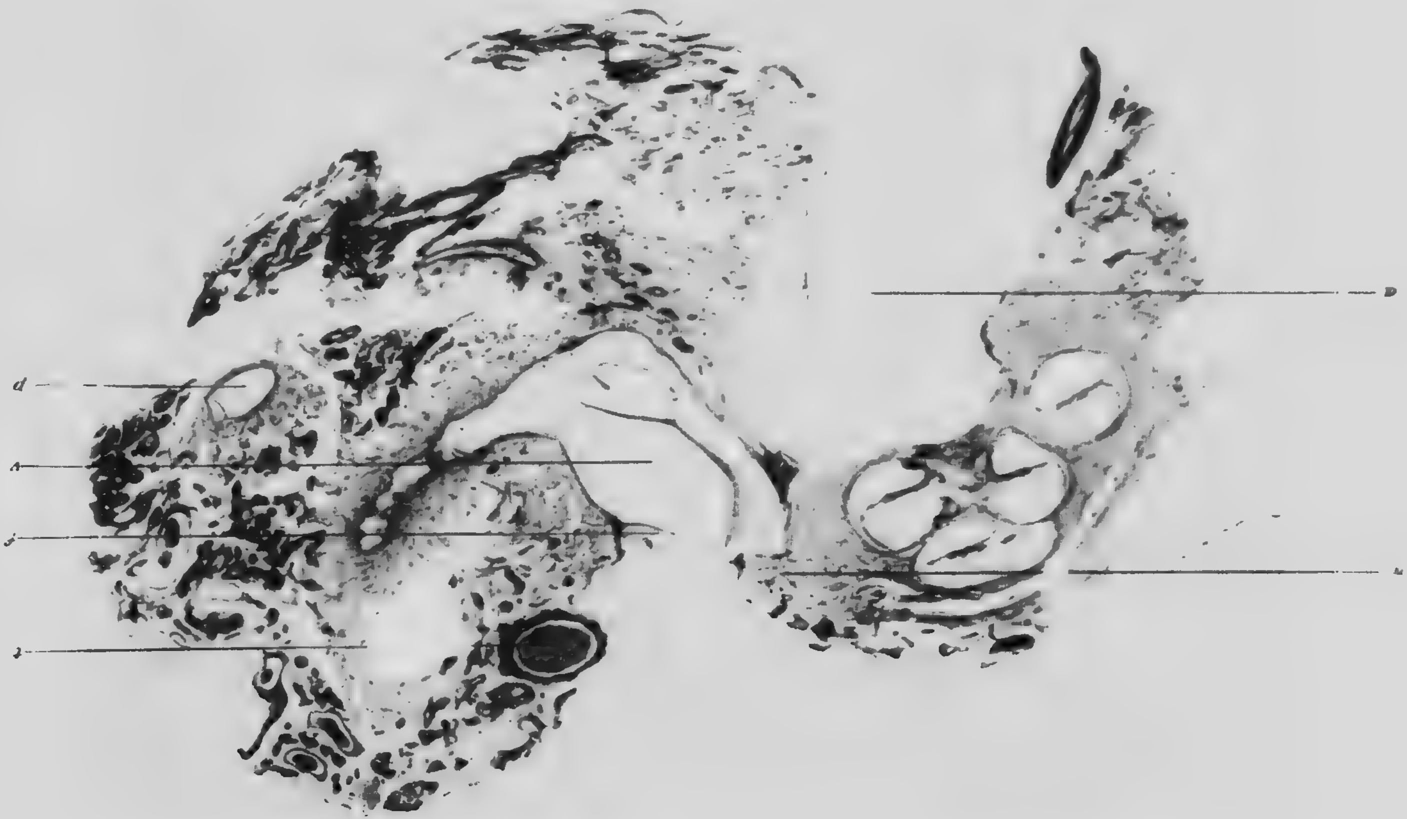
a. Interi[illegible]

β. [illegible]

γ. [illegible]

δ. [illegible]

ε. [illegible]


d
h
j
z
D
k

brane or to the organ of Corti. The organ of Corti presents perfectly normal appearances, the arch is well formed, the rods are present and natural in position, and although the hair-cells are swollen and ill-defined owing to post-mortem disintegration, the hairs themselves are present.

The cells of the ganglion spirale are normal, as also are the other nerve structures.

The tensor tympani and stapedius muscles are quite normal in appearance.

Left ear.—Microscopic examination of the left ear reveals a condition almost identical with that found on the right side. This is in keeping with what is well known in regard to the symmetrical character of the disease.

The position of the diseased area in the bone is immediately in front of the oval window, and in shape and texture is exactly similar to that already described on the right side. On its inner aspect the affected area of bone is limited by the endosteum of the vestibule, and on its outer aspect by the muco-periosteum of the tympanum. Posteriorly it is limited by the stapedio-vestibular articulation, and anteriorly it is bounded by a sharp line of demarcation from the normal bony capsule of the labyrinth.

The diseased bone stains less deeply than the surrounding normal bone of the capsule, and the large spaces in the diseased bone are quite filled up with blood-vessels. There is no sign of any deposit of new-formed bone.

Examination with the high power reveals a condition almost exactly similar to that described on the right side. The affected area is highly vascular. The osteoblasts are rounder in shape than in normal bone, and the canaliculi are few and short.

There is considerable activity in some of the islands of cartilage which are frequently present under normal circumstances in the bony capsule. Some of the cartilage cells are clearly in a state of degeneration, and even disintegration, while in others rapid cell division is taking place. It is interesting to note that in one of these cartilage cells, nuclear division by karyokinesis is present.

The tensor tympani and stapedius muscles are perfectly normal in appearance.

Membranous Structures.—On the left side, as previously described on the right side, no definite pathological change is to be found in the membranous labyrinth. The cells of the organ of Corti are seen to be normal, excepting in respect to such changes as may clearly be considered post-mortem. The stria vascularis is normal, there is no depression or bulging of the membrane of Reissner beyond such as would be expected to occur in preparation.

The cells of the ganglion spirale are healthy, as also are the nerve-fibres running into and out from the ganglion.

CASE III (FIGS. 9-14)

Clinical Examination.--Miss J. D., housekeeper, æt. 55. Examined by myself in Glasgow Cancer Hospital, Jan. 20th, 1914.

The patient has been deaf in both ears for more than twenty-five years. A few years after the onset of the deafness she began to suffer from tinnitus, a symptom which has remained since. It is severe, and causes her considerable distress. In character she likens it to the hissing of steam. She has never noticed the occurrence of paracusis.

Family History.—The grandfather was deaf, but this may have been from old age. One aunt on the father's side was deaf, and in her case the deafness was not due to old age. There is no other case of deafness known among the relatives.

Examination.—Both membranes are noticeably indrawn. No rosy tint is present in the tympanic image, and there is no discharge from the ear. There is the appearance of an old scar in the right membrane.

Right Ear.—A tuning fork (440 vibs.) is not heard in the right ear by air conduction, and only for 7 seconds by bone conduction. (Normal is 17 secs. by bone conduction for this particular fork.)

Whisper: Not heard at all.
Watch: Not heard at all.
Conv. voice: Not heard at all.

Galton's whistle: A considerable portion of the upper range is lost to hearing (Mdh. 6·0, Pfl. 5·8).

Four months after the clinical examination was made, the patient died as a result of secondary malignant deposits in the lungs. A post-mortem was made about twenty-four hours after death, and the temporal bones were removed and prepared for cutting by the method described.

Pathological Examination: *Macroscopic Appearances.*—*Right Ear.*—On examination it was found that there was clear evidence of a former perforation of the membrane in the posterior inferior quadrant. This had been closed over by scar tissue. The middle ear showed no evidence of disease, the mucous membrane in all parts presenting a perfectly normal appearance. The Eustachian tube was freely permeable.

The three ossicles seemed to be free of disease so far as the naked-eye appearances went. There was no evidence of fixation of the stapes in the oval window so far as could be ascertained, but there was not much significance in this, as no attempt was made by pressure to find out the mobility of the ossicles, for fear of damaging the preparation. (It will be seen later that the stapes was fixed by bony ankylosis.)

The petrous portion of the bone was prepared for microscopic examination in the manner previously described, and a complete series of sections was made as nearly as could be arranged in the plane of the modiolus of the cochlea.

Microscopic Examination.—A diseased focus of bone is found in the usual situation in cases of otosclerosis, immediately in front of the oval window. The diseased area is oval in horizontal section and extends from the endosteum of the cochlea to the muco-periosteum of the tympanum. Posteriorly at its greatest extent it reaches just beyond the stapedio-vestibular articulation, and into the foot-plate of the stapes, thus causing ankylosis.

It stains hardly any deeper than the normal bone of the labyrinthine capsule and is only distinguishable from the latter by its more porous character and the difference in shape of the osteoblasts. The absence of any strong affinity for absorbing the stain may be due to one of two causes, either that the process of new bone formation has been extremely slow, or, that it took place many years ago and has long been inactive. Either of these explanations is compatible with the long duration of the symptoms (twenty-five years).

The diseased focus extends downwards into the cleft between the round window and the second whorl of the cochlea.

The line of demarcation is sharply defined, and in some sections of the series blood-vessels may be seen passing out from the diseased focus through the line of demarcation and into the surrounding normal bone.

The spaces within the focus of the diseased bone are filled with marrow, and in many of them blood vessels may also be seen. The bony tissue itself is well lamellated. The actual bone-cells in the diseased area, however, differ to a certain extent from the cells of the normal bone. They are less regular in contour, and the spindle shape so characteristic of normal osteoblasts is not so pronounced. Consequently the diseased bone presents a rather more granular appearance than the surrounding normal bone. No osteoclasts are found.

At one point, as stated above, the new-formed bone has extended across the stapedio-vestibular articulation and into the adjacent region of the foot-plate of the stapes. The bridge of bone thus formed is very narrow, and in depth it only extends through three, or at most four consecutive sections of the series. When it is remembered that the disease had been in existence for twenty-five years, it is surprising that the anatomical change at this point is so small in extent. I have referred to this feature of otosclerosis previously.[1]

In certain regions there is considerable activity in the groups of cartilage cells which are normally found scattered about the neighbourhood of the capsule of the labyrinth.

There is no other focus of diseased bone to be found in the labyrinth.

As regards the membranous structures, it may be said in general that no sign of pathological change could be found anywhere. The organ of Corti, the nerve fibres, the tectorial membrane, and the membrane of Reissner are all normal in structure and position. Finally, in respect to the nerve cells of the ganglion spirale, it may be said that no pathological change is discoverable though of course there are present the usual changes which result from post-mortem disintegration. The nerve fibres are normal. It must again be emphasised, however, that although no pathological changes are found in the ganglion spirale, it must not be assumed that no such changes exist; for the present methods of microscopic examination of the nervous system are far too crude to elucidate changes which we know, on other grounds, must be present. Thus, in this case we know that severe tinnitus was complained of for nearly twenty years, and it is inconceivable that such a symptom could exist for that period of time without being associated in some way, either of cause or effect, with changes in the nerve cells in some part of the auditory tract, and probably in all.

---

[1] Gray, *Diseases of the Ear*, p. 315 (Baillière, Tindall & Cox, 1910).

Left Ear.—Family history, subjective symptoms, etc., as recorded above in the clinical report of the right ear.

Clinical Examination.—Membrane markedly indrawn. No rosy tint present, and no evidence of any previous suppurative disease.

Tuning fork (440 vibs.) not heard at all by air conduction; heard 5 secs. by bone conduction.

Whisper: Not heard at all.

Conv. voice: Just heard within one inch of the ear.

Edelmann-Galton whistle: A considerable number of notes lost at the upper end of the scale (Mdh. 6·0, Pfl. 5·8).

Pathological Examination: *Macroscopic.*—The temporal bone was prepared by the method described above.

Though the tympanic membrane is indrawn, there is no sign of structural alteration visible to the naked eye.

The mucous membrane of the middle ear presents a perfectly healthy appearance; and the Eustachian tube, in so far as it is present, is unobstructed. But it must be noted that the naso-pharyngeal portion of the tube was left behind in the cadaver, and there may, therefore, have been some pathological change in it or in the naso-pharynx itself.

The veins of the promontory are somewhat dilated, and some bulging exostoses are present in the same region. Immediately below the oval window there is seen to be a peculiar spurlike exostosis.

CASE III—FIG. 9

RIGHT EAR

HORIZONTAL SECTION THROUGH THE PETROUS PORTION OF THE TEMPORAL BONE IN THE PLANE OF THE STAPES. × 8

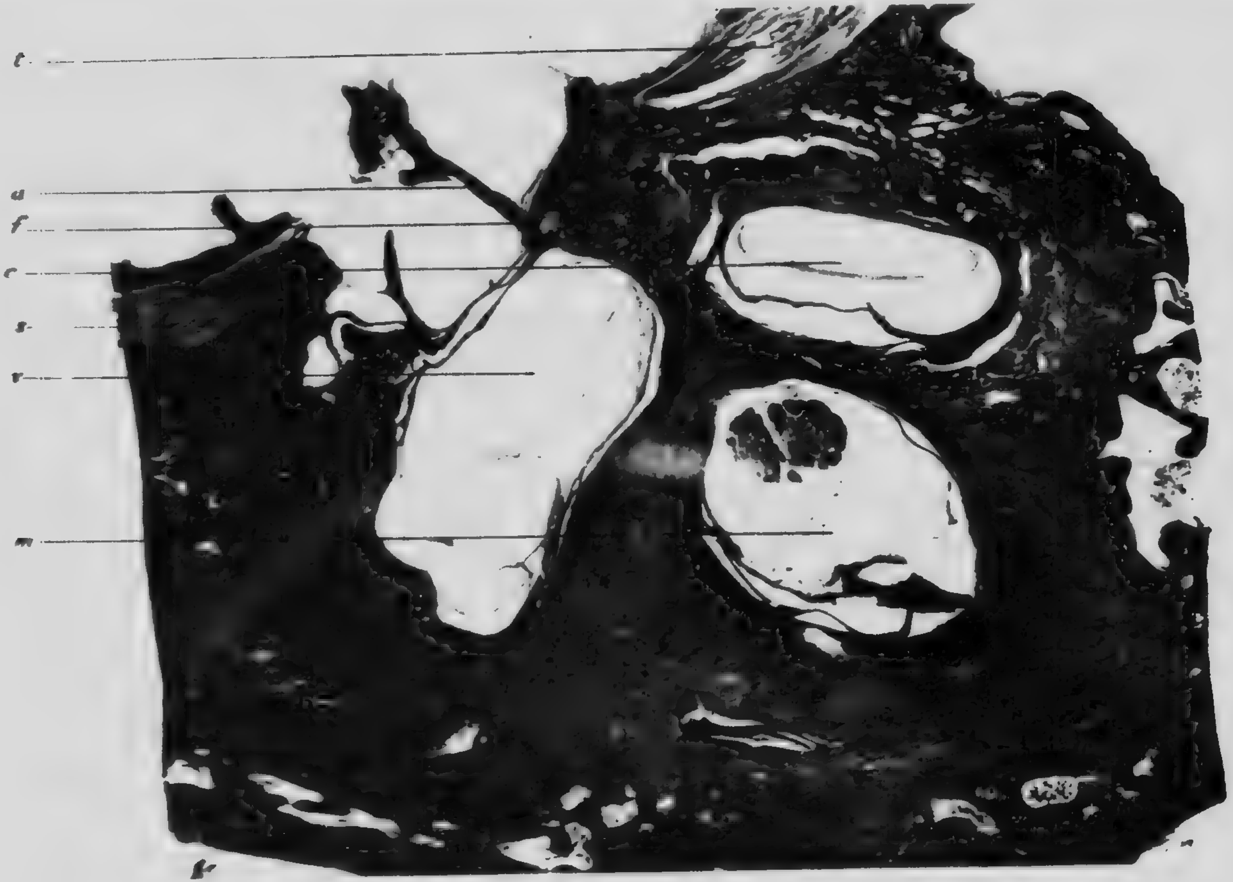
t. Tensor tympani muscle.
a. Anterior crus of stapes.
f. Focus of rarefied diseased bone. It is not easily distinguishable from the surrounding bone, and for that reason, as well as from the clinical history, the inference may be drawn that in all probability it is not of recent origin. The focus is seen more clearly in fig. 10.
c. Tangential section through one of the whorls of the cochlea.
s. Stapedius muscle.
v. Vestibule.
m. Internal auditory meatus.

[illegible]

[illegible]

HORIZONTAL SECTION [illegible]
[illegible]

t. Tensor [illegible]
a. Ant[illegible]
f. Focus [illegible] the [illegible] patient, [illegible]
c. Tan[illegible]
s. Stap[illegible]
v. Vestibul[illegible]
m. [illegible]


t
a
f
c
s
r
m
k

CASE III—FIG. 10

RIGHT EAR

HORIZONTAL SECTION THROUGH THE FOCUS OF DISEASED BONE IN FRONT OF THE OVAL WINDOW. × 60 ca.

f, f, f. Focus of diseased porous bone. In the spaces marrow and blood-vessels may be seen. The diseased bone is well laminated. It stains hardly any more deeply than the old normal bone, doubtless for the reason that it is not of recent formation.

d, d, d. Line of demarcation. In some parts it is very easily distinguished, while at others it is only just visible, the diseased bone and the old normal bone appearing to differ but slightly in texture.

o, o. Old normal bone of the capsule of the labyrinth.

v. Vestibule.

a. Annular ligament.

b. Bridge of diseased bone uniting the footplate of the stapes with the wall of the oval window. (See fig. 11.)

m. Mucous lining of the tympanum.

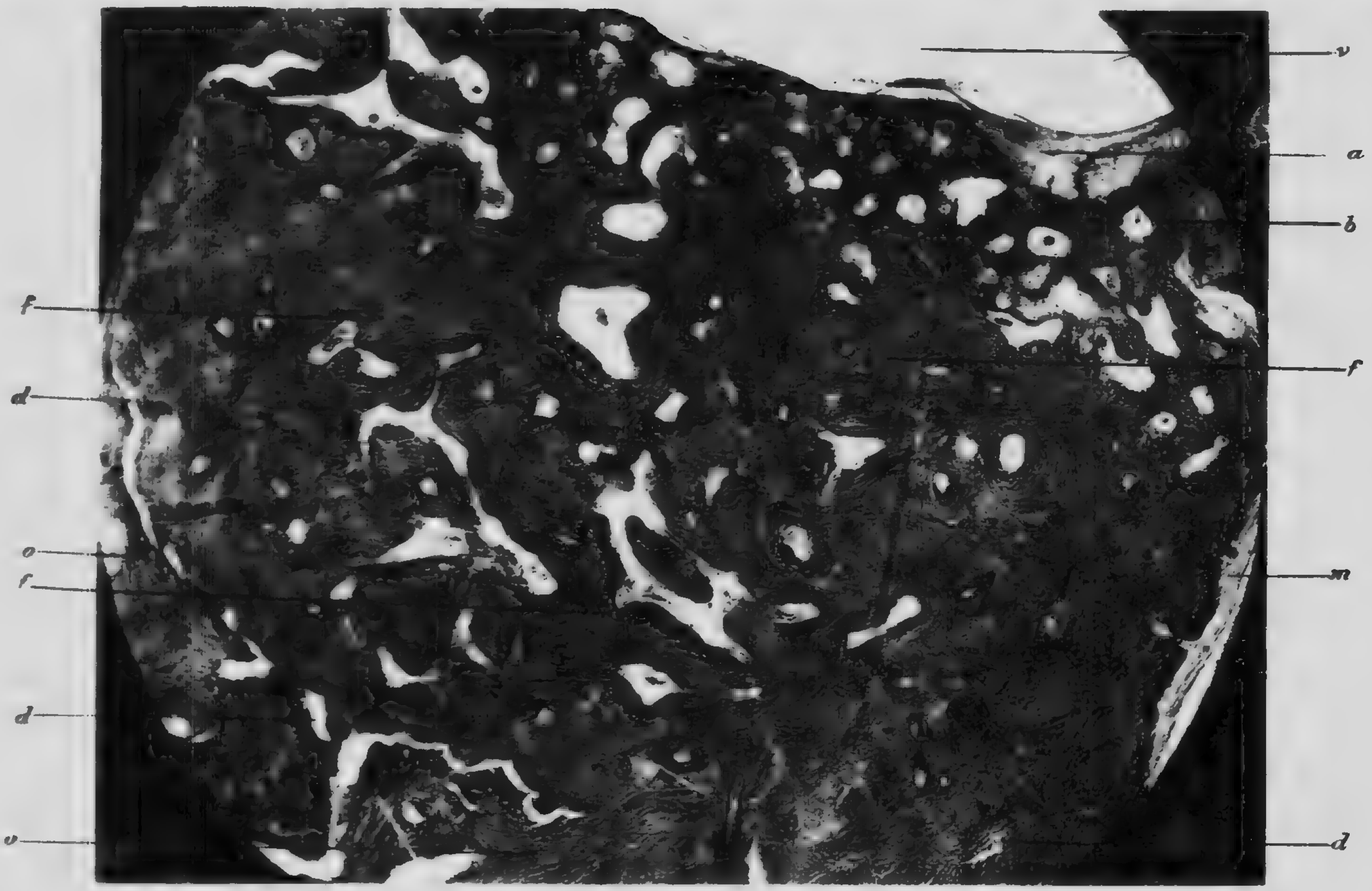
[illegible]

HORI[illegible] IN [illegible]

[illegible] and [illegible]

[illegible] slightly [illegible]

[illegible]


f
d
o
r
d
v
v
a
b
f
m
d

CASE III—FIG. 11

RIGHT EAR

HORIZONTAL SECTION THROUGH THE STAPEDIO-VESTIBULAR ARTICULATION AND THE ADJACENT PORTIONS OF THE STAPES AND VESTIBULAR WALL OF THE OVAL WINDOW. THE WHOLE OF THE RIGHT OF THE PHOTOGRAPH SHOWS A PORTION OF THE FOCUS OF DISEASED POROUS BONE, WHILE TO THE LEFT ARE SHOWN PORTIONS OF THE ANTERIOR CRUS AND FOOTPLATE OF THE STAPES. × 120 ca.

m. Mucous membrane of tympanum.
a. Anterior crus of stapes.
b. Bridge of bone passing from the wall of the fenestra ovalis to the footplate of the stapes and causing ankylosis. The bridge is very thin and only occupied a depth of three or four thin consecutive sections cut by the microtome.
f. Footplate of stapes.
n. Focus of diseased porous bone.
v. Vestibule.

[illegible]

[illegible]

HORSE'S [illegible] ARTICULATION [illegible] THE [illegible] WALL [illegible] THE [illegible] THE [illegible] SHOWING PORTIONS OF THE [illegible] BONE, [illegible] THE LEFT [illegible] THE ANTERIOR [illegible] AND POSTERIOR [illegible] 12 [illegible]

m. Ma[illegible]

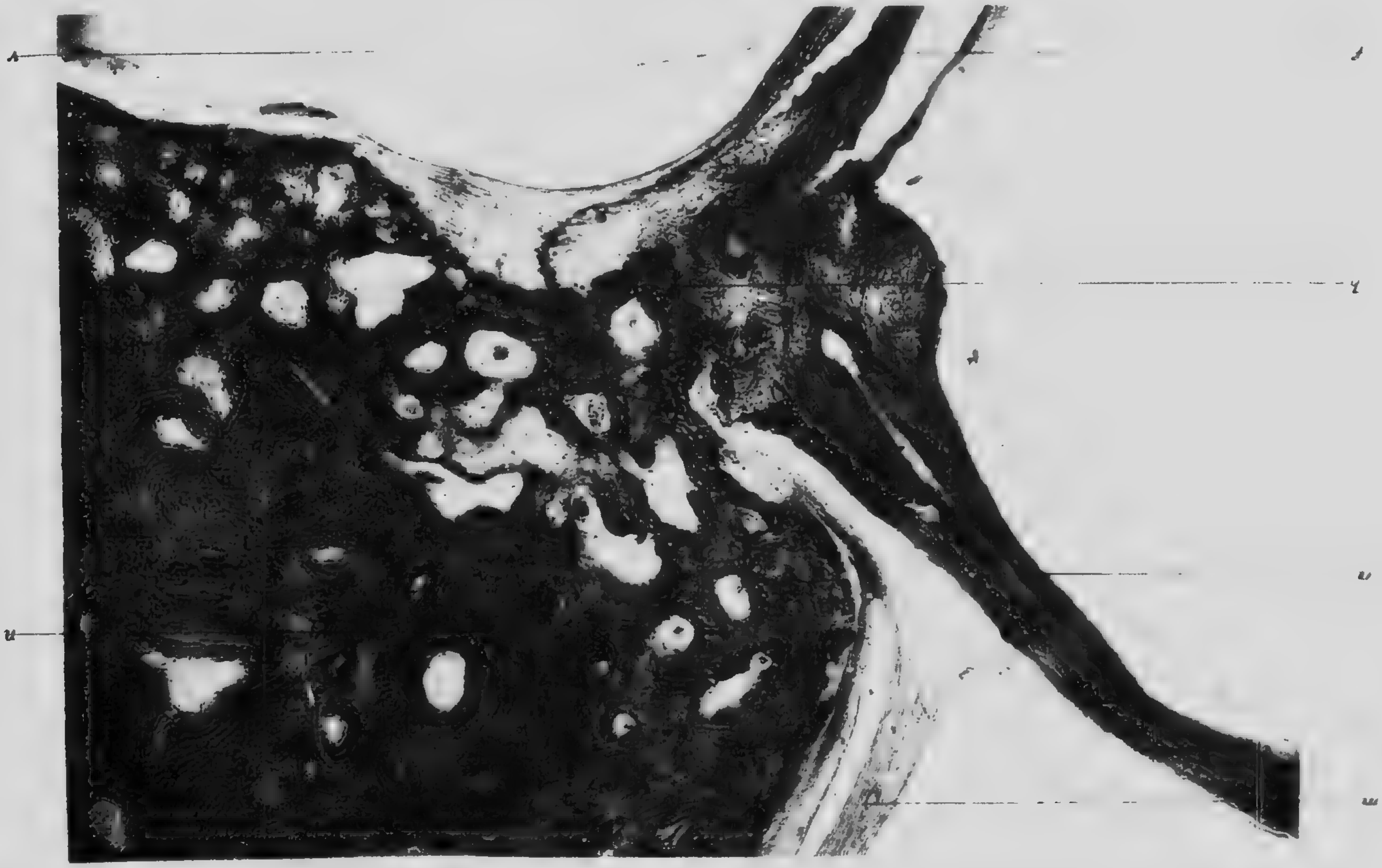
a. Ante[illegible]

b. Bridge [illegible] to the [illegible] is [illegible]

f [illegible]

e [illegible]

p [illegible]

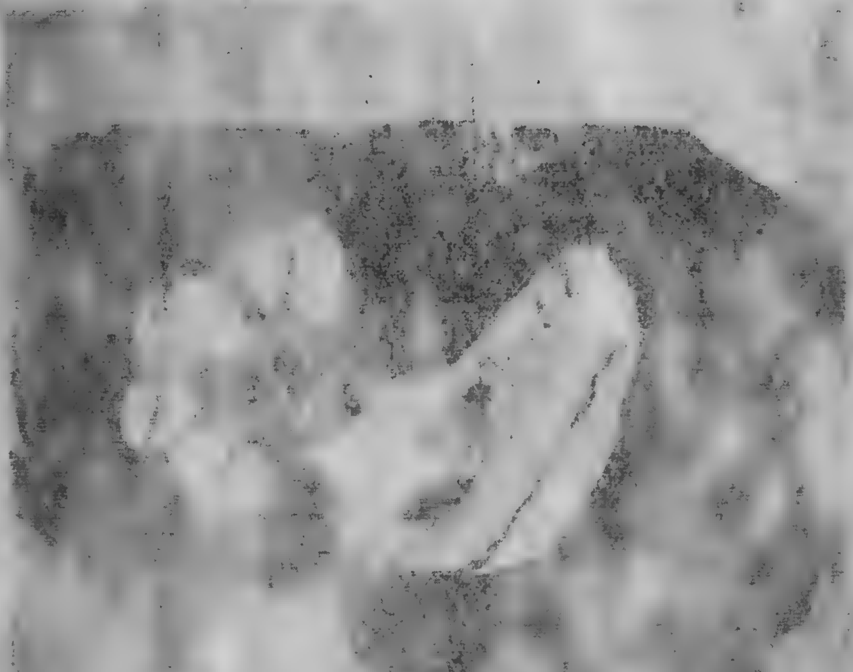

s
a
h
f
q
n
m

CASE III—FIG. 12

LEFT EAR

HORIZONTAL SECTION THROUGH THE LABYRINTH. × 5

f. **Facial nerve.**
v. **Vestibule.**
m. **Internal auditory meatus.**
n. **Focus of diseased bone. It will be observed that the diseased bone does not stain any more deeply than the surrounding bone. In this respect the case contrasts sharply with Case I, the reason of the difference being probably the difference in duration of the disease.**

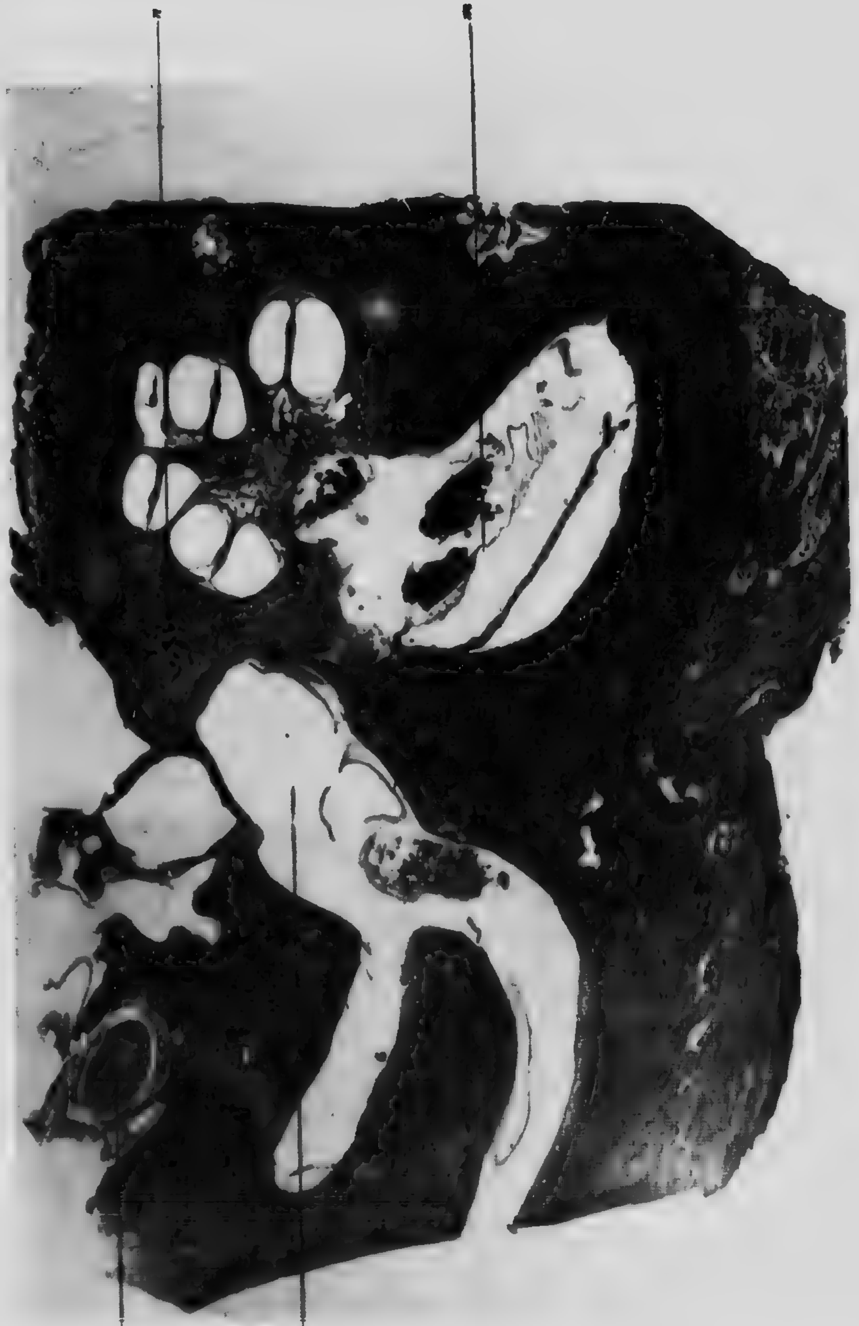


[illegible]

LEFT EAR

HORIZONTAL SECTION THROUGH THE [illegible]

- *f.* **Facial** nerve.
- *v.* **Vestibule.**
- *m.* **Internal** auditory meatus.
- *n.* **Focus** of diseased bone. It will be observed that the diseased bone is not stained more deeply than the surrounding bone. In this respect the case contrasts sharply with those [illegible]



Case III—Fig. 13

LEFT EAR

Horizontal Section showing the Focus of Diseased Bone

× 60 ca.

d, d, d. Line of demarcation. It is to be noted that the focus of diseased bone is characterised by the large number of spaces filled with marrow and blood-vessels. It does not, however, stain any more deeply than the old normal bone of the capsule outside the line of demarcation. It has, however, a rather more granular texture owing to the fact that the osteoblasts are not spindle-shaped as are those in the normal bony capsule.

o. Normal bone of the capsule.

n. Focus of diseased bone.

t. Muco-periosteum of the tympanum.

s. Anterior extremity of footplate of stapes.

v. Vestibule.

PLATE III. FIG. 4.

LEFT EAR.

HORIZONTAL SECTION SHOWING THE FOCUS OF DISEASED BONE
× [illegible].

d, d. Line of demarcation. It is to be noted that the focus of diseased bone is characterised by the large [illegible] of spaces filled with marrow and blood-vessels. It does not, however, stain any more deeply than the old normal bone of the capsule outside the line of demarcation. It has, however, a rather more granular texture owing to the fact that the osteoblasts are not spindle shaped as are those in the normal bony capsule.

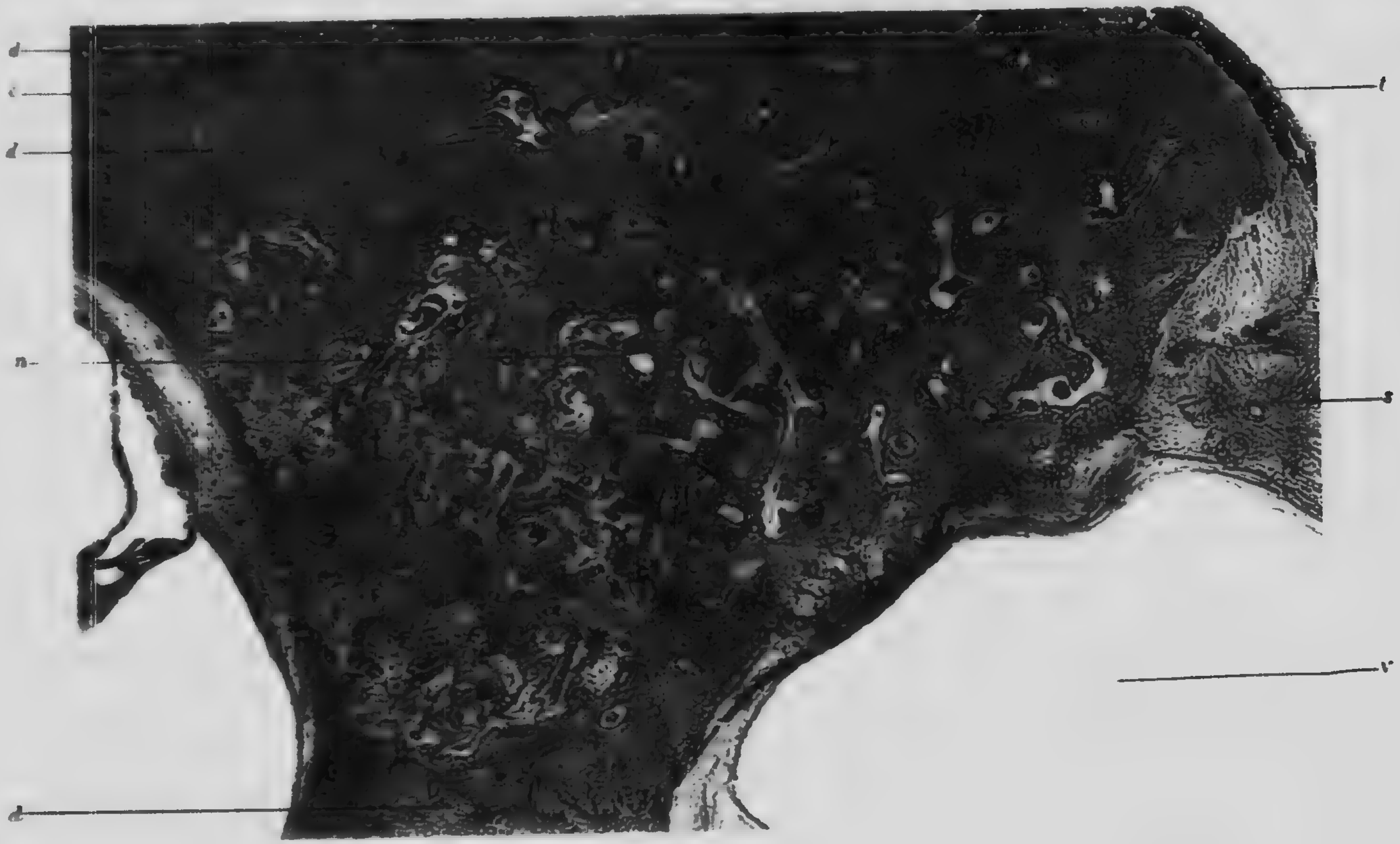
o. Normal bone of the capsule.

n. Focus of diseased bone.

l. Mucous periosteum of the tympanum.

s. Anterior extremity of footplate of stapes.

v. Vestibule.


d
c
d
n
d
t
s
v

CASE III—FIG. 14

LEFT EAR

HORIZONTAL SECTION THROUGH THE STAPEDIO-VESTIBULAR ARTICULATION. × 180 ca.

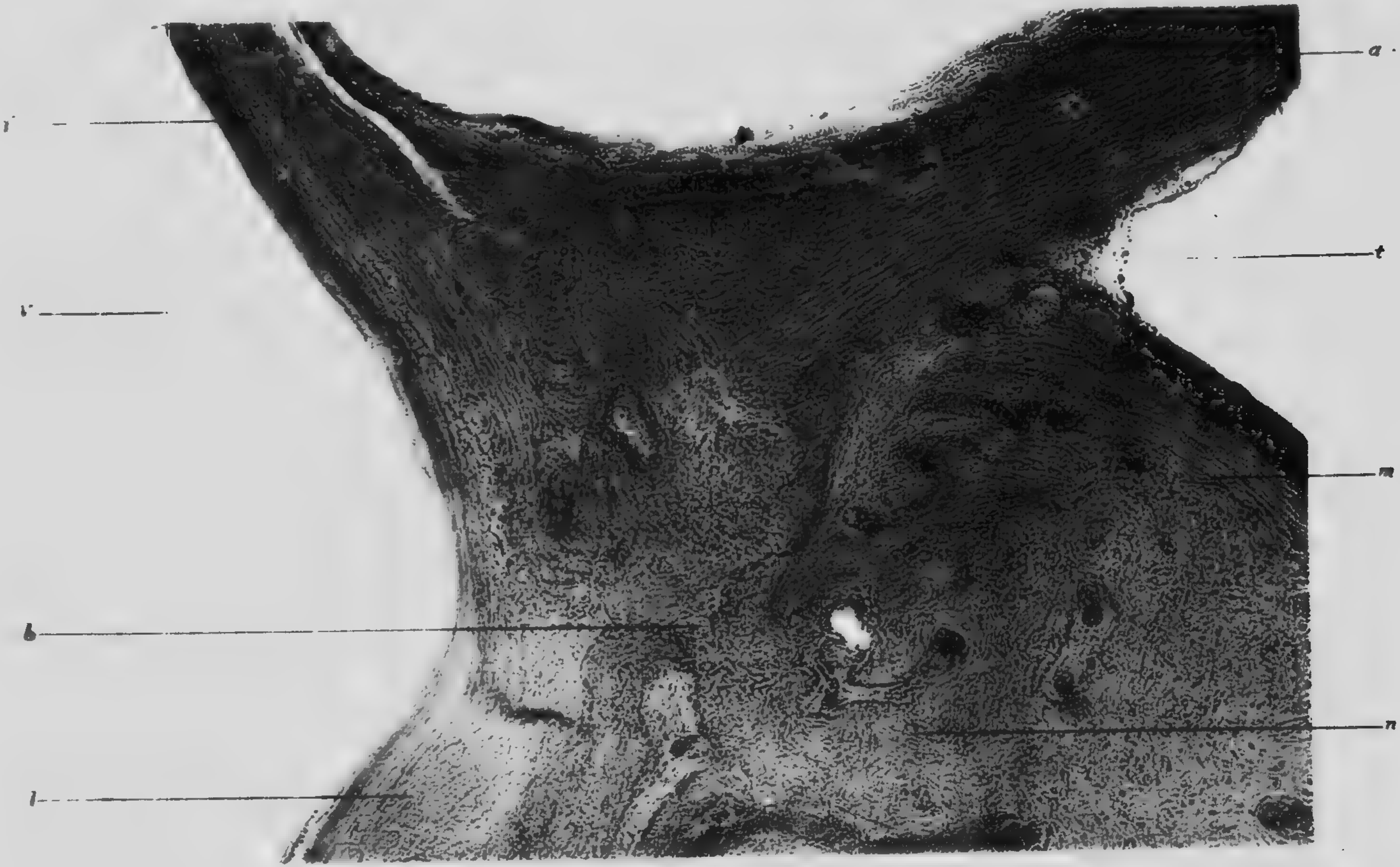
f. Footplate of stapes.
v. Vestibule.
b. Bridge of bone uniting the footplate of the stapes with the wall of the oval window.
l. Annular ligament.
n. Focus of diseased bone.
m. Muco-periosteum of tympanum.
t. Tympanic cavity.
a. Anterior crus of stapes.

CASE [illegible] FIG. [illegible]

LEFT EAR

HORIZONTAL SECTION THROUGH THE STAPEDIO-VESTIBULAR ARTICULATION. × 180 ca.

- **f.** Footplate of stapes.
- **v.** Vestibule.
- **b.** Bridge of bone uniting the footplate of the stapes with the wall of the oval window.
- **l.** Annular ligament.
- **n.** Focus of diseased bone.
- **m.** Muco-periosteum of [illegible]
- **t.** Tympanic cavity.
- **a.** Anterior crus of stapes.


a
t
m
n
l'
b'
b
l

All three ossicles are quite normal in appearance. The stapes does not show any evidence of fixation, but as will be seen later, when examined by the microscope it was found that as a matter of fact the bonelet was fixed by osseous ankylosis.

Microscopic Examination.—Sections were cut in the horizontal plane, and it may be said at once that they revealed pathological changes almost identical with those on the right side; indeed, it might almost be said that the left side was a replica of the right. This remarkable symmetry has also been recorded in Cases I and II, and probably would have been found to exist also in Case IV, had both temporal bones been sent for examination.

For this reason, therefore, it is not necessary to enter into such detail in regard to the pathological changes on the left side as on the right.

It will be observed (fig. 12) that the focus of diseased bone occupies the usual position immediately in front of the oval window, and extends from the stapedio-vestibular articulation to the cochlea. Externally it reaches to the muco-periosteum of the tympanum, and internally to the vestibule. There is no other focus of diseased bone.

The footplate of the stapes is united to the wall of the oval window by a narrow bridge of bone, but this is only found in three or four consecutive sections of the series, and is therefore very thin in its vertical diameter. The muco-periosteum of the tympanum is perfectly healthy in appearance. The affected area of bone is sharply delimited from the surrounding normal bone, but in certain regions the line of demarcation is difficult to find because the staining property of the two different types of bone hardly varies. In other parts of its course the line of demarcation is easily recognised. The texture of the affected bone is very different from the adjacent normal bone. There are many spaces in the bone which are filled with blood-vessels and marrow, and the osteoblasts are rounder and less markedly spindle-shaped than in the normal bone. Careful search for osteoclasts fails to reveal their presence, and indeed the whole appearance of the bone both at the line of demarcation and elsewhere gives the impression that no special activity is present beyond that which would be found in normal bony tissue. In other words, the cellular activity in the bone has long since passed away. There is no evidence whatever of inflammatory activity either past or present.

The tensor tympani and stapedius muscle are both perfectly normal in appearance, there being no sign of fatty or other degeneration.

The membranous structures of the inner ear show no sign of pathological changes, though of course there are the usual artefacts due to post-mortem disintegration and methods of preparation.

The ganglion spirale shows no sign of pathological change, though very possibly such changes are really present, but are not discoverable by any of the present microscopical methods applicable to the labyrinth.

CASE IV (FIGS. 15-20)

In the following case no clinical examination was made. The patient, some time before her death, indicated that she would like to have an examination made of her ear, in order that an attempt might be made to find out the cause of her deafness, and add to our knowledge of such conditions. At the post-mortem examination, only the left temporal bone was removed. I have to thank Mr. Tilley for obtaining this specimen for me. The patient's death occurred at the age of 85, the symptoms having existed for about sixty years.

The history of the case was to this effect. The patient, Miss H., at the age of about 20, had become dull of hearing fairly quickly and without apparent cause. Ultimately the deafness reached a high degree, until she was unable to hear the voice at all in the left ear, and was compelled to use an ear trumpet in the right ear. Noises in the ear were more or less constantly present, but not to any distressing degree. The patient had never complained of giddiness. There was a marked tendency to deafness in the patient's family and ancestors.

Pathological Examination.—Before the preparation reached me it had been preserved for a considerable time in rather weak spirit, and this accounts in part for the fact that the more delicate structures of the organ of Corti, etc., are not shown with the same clearness of definition as in the previous case.

The external meatus is, perhaps, somewhat narrower than normal, but not to such an extent as to be considered pathological. There is no cerumen in the meatus. The tympanic membrane is normal in position, and there is no perforation or scar. The Eustachian tube is healthy throughout all the portion present in the preparation; the naso-pharyngeal portion is not included, so that its condition cannot be ascertained. There is no fluid or discharge of any kind present in the tympanum, there is no visible sign of pathological change in the mucous membrane and no adhesions are present. The two outer ossicles appear to be healthy, as also does the tensor tympani muscle so far as the naked eye can ascertain.

The stapes is ankylosed in the oval window, and its anterior crus is firmly welded by bone with the inferior anterior portion of the pelvis of the oval window. The round window is considerably narrowed by the marked hyperostosis of the bone which forms its lip. There are one or two minute exostoses immediately in front of the oval window and a little above the promontory.

The internal auditory meatus is distinctly narrowed, and on the posterior margin of the entrance is a marked bulging which further decreases its lumen.

The preparation was decalcified, treated, and stained by the methods described above. A complete series of horizontal sections was made.

A photograph of a section passing through the two crura and the footplate of the stapes is shown in fig. 16. A general view of the whole section shows that the internal auditory meatus is much narrower than in normal circumstances and the vestibule is reduced in size. The footplate of the stapes is very greatly thickened in its anterior portion, and the bony portion of the whole of the footplate is quite altered in character from the normal. It is porous in texture, and in fact is identical with that found in the focus of otosclerotic bone immediately in front of the oval window. This fact is demonstrated more clearly when higher magnifications are employed.

Close examination shows that the footplate is ankylosed both at its anterior and posterior extremities by bridges of bone extending across to the walls of the oval window. The tissues of which the anterior and posterior bridges are composed are, as above stated, identical with otosclerotic bony tissue; indeed, they are parts of one large otosclerotic focus extending from a line a short distance in front of the anterior margin of the oval window to a line a short distance behind its posterior margin.

There is, however, another and different change in the bone in this case. This consists of a comparatively large area of highly rarefied bone in the region lying between the posterior margin of the oval window, the facial nerve, and the horizontal semicircular canal.

In this case, therefore, there are two distinctly different changes in the bone. This statement must not be taken to mean that the cellular changes are pathologically fundamentally different, or arise from different causes. It means that, whatever the pathological process or processes may be, the anatomical result is very different in the two regions described.

Taking first the ordinary otosclerotic change which extends anteriorly from the line d, d in fig. 17, to the line d, d in fig. 18, there is not very much to describe beyond the fact that the bony tissue resembles that shown in an early stage in Case I, and in a late stage in Case III. The new-formed bony tissue stains rather more deeply than the adjacent normal bone, and is marked off by the usual sharp line of demarcation. The marrow spaces in the diseased porous bone are rather small in comparison with those in Case III, and very small in comparison with those in Case I. This is what is to be expected considering the duration of the disease in the three cases. The bony tissue of the diseased area is perfectly well lamellated. As is usually the case, the osteoblasts are not quite so noticeably spindle-shaped as in the normal bone, being rounder or more irregular in shape. There is no round-cell infiltration, nor any other sign of inflammatory activity. Osteoclasts are not found.

The mucous membrane in the cleft between the footplate of the stapes and the wall of the basin of the oval window shows a certain amount of fibrous thickening, but there is no round-cell infiltration. A few irregular structures are seen scattered about in the mucous lining of this region, but they do not appear to be cellular, and very possibly indicate the position of calcareous deposits, though, of course, the calcareous salts have been removed during decalcification. Similar structures are seen in the thickened mucous lining on the outer aspect of the posterior position of the footplate of the stapes. The rest of the mucous lining of the tympanum is normal in appearance.

The other type of anatomical change in the bone is that shown in figs. 16, 19 and 20 in the region lying between the posterior margin of the oval window, the facial nerve, and the horizontal semicircular canal. This type has never hitherto been described, so far as the writer is aware. It consists of tissue in which the mineral element is reduced by the process of rarefication to such an extreme degree that the marrow spaces cover a much larger surface than the bony element. Indeed, the latter is reduced to a few fine spicules and trabeculæ. In regard to this area of diseased bone, it is very interesting to note that, although the process of absorption of bone has been carried out to such a great degree, yet the deposition of bone is by no means absent. New bone has been deposited; and, if the intensity of the stain affords any clue to the age of the bone, then it may be inferred that the new-formed bone was in process of deposition up to a period very shortly before the death of the patient.

The newly deposited bone in this area forms a lining along the edge of the old normal bone which is being absorbed. The new-formed bone is quite well lamellated, and, indeed, is only differentiated from the old normal bone by its deeper staining capacity and the sharp line of demarcation. It is important to observe that the line of demarcation is as sharp as that found in the ordinary otosclerotic change seen in front of the oval window. There is no round-cell infiltration nor any sign of inflammatory activity. In this diseased area there are certain points at which the layer of dark-stained new bone, lining the outside of the old normal bone, is incomplete, so that the old normal bone lies quite bare to the marrow spaces. This is shown in fig. 20, in which there may be seen an osteoclast which has been fixed in the process of eating up both the new-formed bone and the old normal bone. In the same figure several osteoblasts may be seen escaping from their calcareous surrounding and apparently coalescing to form osteoclasts in the manner described by Kölliker. This is an important fact, because Manasse maintains that in otosclerosis the process of absorption of the bone is carried out, not by osteoclasts, but by simple pressure. Now, whether Manasse is or is not right in his view regarding the bony change most common in otosclerosis, his view cannot be held to be correct in regard to the bony change in the diseased area under consideration.

My own view of the matter is that the processes seen in the two regions in this case are fundamentally similar, though the anatomical result is so different. That is to say, they both consist first in the constant slow absorption of bone followed immediately by the slow deposition of bone. The different anatomical result, then, would depend merely on the relative rates of these two factors. If the process of deposition occurs more extensively than that of absorption, as is usually the case, then there is an increase in the total amount of bone, and the new-formed focus of bone tends to become dense and also to occupy a larger area than before, thus causing exostosis and hyperostosis and narrowing of the round and oval windows. This is the type of new bone formation associated in the mind of aurists with otosclerosis.

If, on the other hand, the process of deposition proceeds more slowly than absorption, then the bone undergoes a constantly increasing process of rarefaction, and does not increase in volume. This is what is found in the area of highly rarefied bone found in the case under discussion, and a somewhat similar process is apparent in Case II, though in the latter there is not to be found any deposit of new bone at all. This type of change in the bone has not hitherto been described.

The reason for the co-existence of these two anatomically different types of diseased bone in the same individual and in close juxtaposition is a matter for conjecture. I am inclined to think that in this particular case the age of the patient at the time when the bony change took place most probably accounts for the difference. The alteration in the bone which has hitherto been regarded as characteristic of otosclerosis, and which is present in this case in front of the oval window and along the whole of the footplate of the stapes, began in comparatively early life, the symptoms dating from about the age of 20. This is confirmed by the fact that when examined by the microscope it is clear that the cellular activities within the focus are little, if at all, more pronounced than in normal bony tissue. For example, osteoclasts are either scanty or absent from many sections, and the bone stains hardly more deeply than the normal surrounding bone. On the other hand, in the region of rarefied bone in front of the facial nerve and the horizontal semicircular canal, a very different state of affairs was clearly in existence at the time of death. Cellular activity was very great, as shown by several facts. For example, the new-formed bone stains deeply, osteoclasts are present in large numbers, and many osteoblasts are seen emerging from their calcareous matrix and coalescing to form osteoclasts.

In this case, therefore, it may be conjectured, with a fair degree of probability, that the different ages at which the disease processes were active account for the difference in the structural results. For, after all, apart from actual pathological changes, it must be remembered that, in so far as changes in bony tissue occur as the result of old age, these changes on the whole are rather of the nature of absorption than of deposition.

Before leaving the subject the reader may be advised to refer again to Case II, in which also the absorption of

9

CASE IV—FIG. 15

LEFT EAR

STEREOSCOPIC VIEW OF THE INNER WALL OF THE TYMPANIC CAVITY, SHOWING HOW THE STAPES IS FIXED

a. Anterior crus of stapes.
s. Tendon of stapedius muscle.
l. Upper margin of the lip of the round window, very much thickened by hyperostosis.
r. Round window, considerably narrowed by hyperostosis.

This photograph should be viewed through a stereoscope.

[illegible]

[illegible]

[illegible]

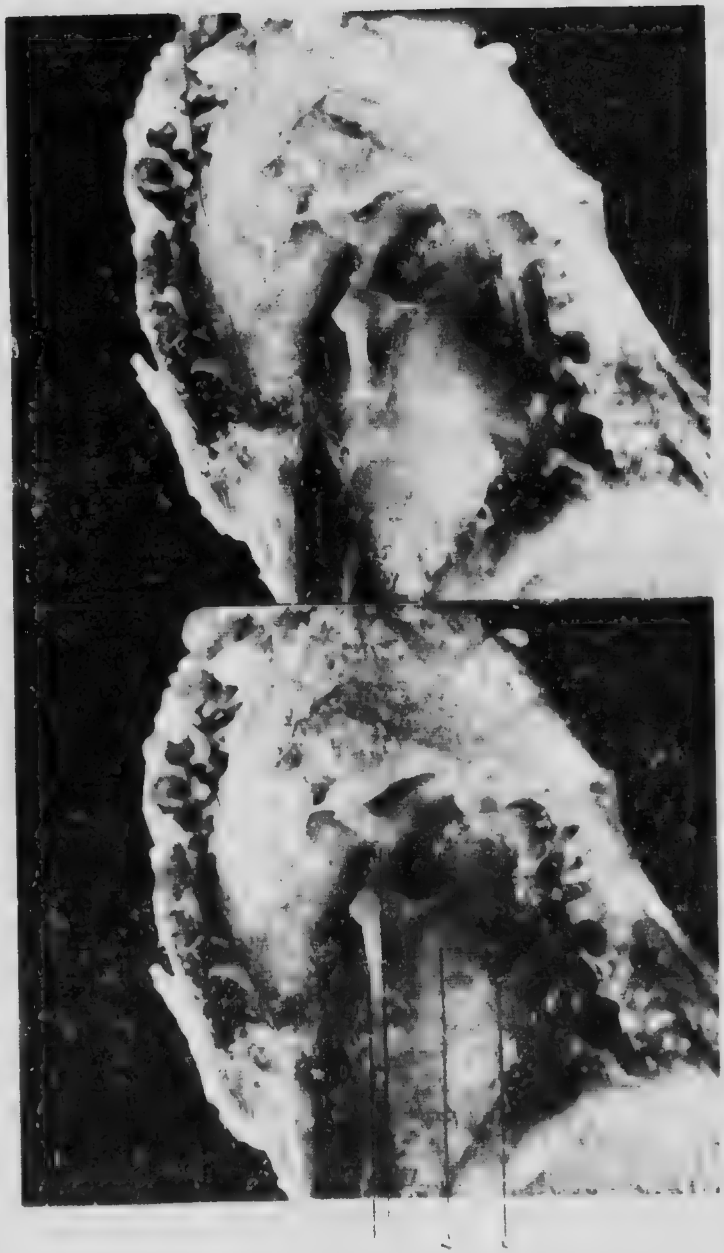
a. A[illegible]

[illegible]

d. [illegible]

e. [illegible]

The [illegible]



Case IV—Fig. 16

LEFT EAR

Horizontal Section through the Petrous Portion of the Temporal Bone. × 8

n. Facial nerve.

r. Large focus of diseased rarefied bone in the region immediately behind the oval window. The process of absorption of bone has been so marked that only a few small islets of bone are seen (darkly stained) surrounded by large spaces filled with marrow. There has also been a process of deposition of new bone in this region, as is shown by the fact that the posterior margin of the footplate of the stapes is united by a bridge of diseased bone to the wall of the oval window.

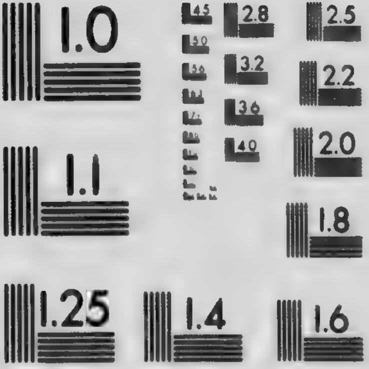
h, h. Horizontal semicircular canal, cut in two different places.

p. Posterior semicircular canal.

f. Footplate of stapes very much thickened by the deposition of diseased cancellous bone. A bridge of the diseased porous bone is seen passing from the anterior margin of the footplate of the stapes to the focus of diseased bone in the region adjacent to the anterior wall of the oval window.

v. Vestibule; considerably smaller than normal.

a. Internal auditory meatus; narrower than normal.



MICROCOPY RESOLUTION TEST CHART
NATIONAL BUREAU OF STANDARDS
STANDARD REFERENCE MATERIAL 1010a
(ANSI and ISO TEST CHART No 2)

a
e
f

CASE IV—FIG. 17

LEFT EAR

HORIZONTAL SECTION THROUGH THE ANTERIOR REGION OF THE STAPEDIO-VESTIBULAR ARTICULATION AND THE ADJACENT PORTIONS OF THE BONY CAPSULE OF THE LABYRINTH AND THE FOOTPLATE OF THE STAPES. × 80 ca.

o, o. Normal bony capsule of the labyrinth.

d, d. Line of demarcation.

n, n. Focus of diseased porous bone, reaching beyond the stapedio-vestibular articulation, into the footplate of the stapes. It will be noticed that the diseased bone is more porous than the normal bone. The lamellation is almost as perfect as in normal bone.

b. Bridge of bone passing from the wall of the oval window to the footplate of the stapes, and causing ankylosis.

f. Footplate of stapes. There is none of the original bony tissue present, the whole of the footplate having been replaced by new-formed porous bone, and at the same time very much thickened.

m, m. Mucous lining of the tympanum passing down into the deep cleft formed by the greatly thickened edges of the footplate of the stapes and the wall of the oval window.

c, c. Irregular masses probably representing calcareous deposits.

v. Vestibule.

i
d
a
c
d
a
c
m
f
c
m
b
v

CASE IV—FIG. 18

LEFT EAR

HORIZONTAL SECTION THROUGH THE POSTERIOR MARGIN OF THE OVAL WINDOW AND ADJACENT STRUCTURES. × 100 ca.

m, m. Large spaces filled with marrow in the highly-rarefied diseased bone. (See fig. 16.)

n. Darkly stained new-formed bone lining the marrow-spaces.

o. Old normal bone of the part. It is gradually being absorbed and the new-formed bone lining the marrow-spaces is deposited in its place.

v. Vestibule.

t. Mucous lining of the tympanum.

b. Bridge of diseased porous bone uniting the posterior margin of the footplate of the stapes with the wall of the oval window.

d, d, d. Line of demarcation between the diseased otosclerotic porous bone and the remains of the old normal bone of this portion of the capsule of the labyrinth.

f. Footplate of the stapes. The normal bone of the footplate has entirely disappeared and been replaced by the porous bony tissue characteristic of otosclerosis.

f
g
p
r
c

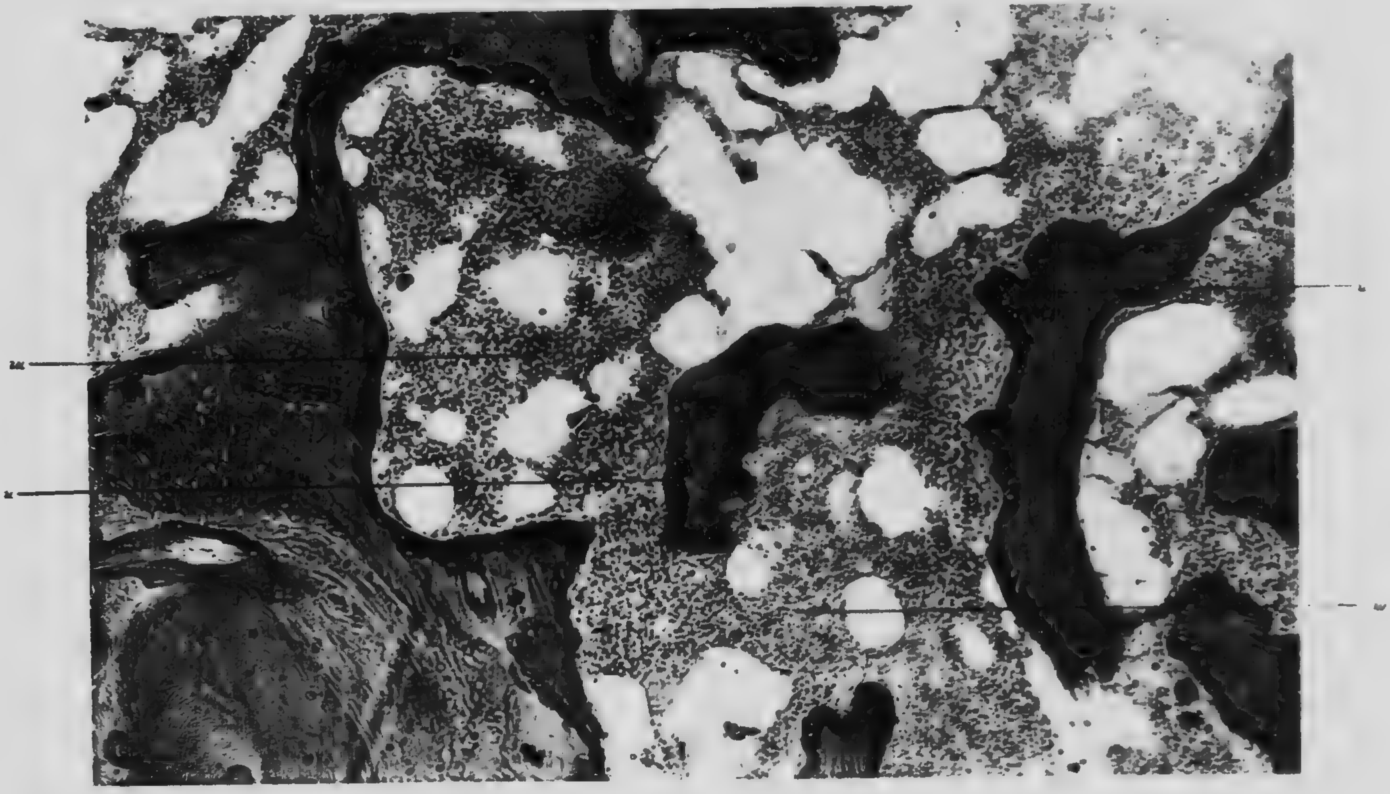
CASE IV—FIG. 19

LEFT EAR

SECTION THROUGH THE REGION ADJACENT TO THE MARGIN AT WHICH THE NORMAL BONE IS BEING TRANSFORMED INTO THE HIGHLY RAREFIED BONE BEHIND THE OVAL WINDOW. THE PORTION TO THE RIGHT AND ABOVE IS THE NORMAL BONE CLOSE TO THE HORIZONTAL SEMICIRCULAR CANAL AS SHOWN IN FIG. 16, AND HAS NO MARROW-SPACE. × 120 ca.

m, *m*. Spaces formed by the absorption of bone and now filled with marrow.

n, *n*. New-formed bone, deeply stained, deposited round the margins of the old bone, faintly stained, as the latter is being absorbed.



CASE IV—FIG. 20

LEFT EAR

SECTION THROUGH A PORTION OF THE HIGHLY RAREFIED BONE IN THE REGION BEHIND THE OVAL WINDOW. × 250 ca.

m. Marrow occupying the space formed by the absorption of bone.
n. Layer of new-formed bone deposited along the outer surface of the old bone as the latter is absorbed.
o. Old normal bone of this region.
p. Phagocytic giant-cell in process of absorbing both the new-formed bone (darkly stained) and the old bone (faintly stained). The cell is an osteoclast.

[illegible]

[illegible]

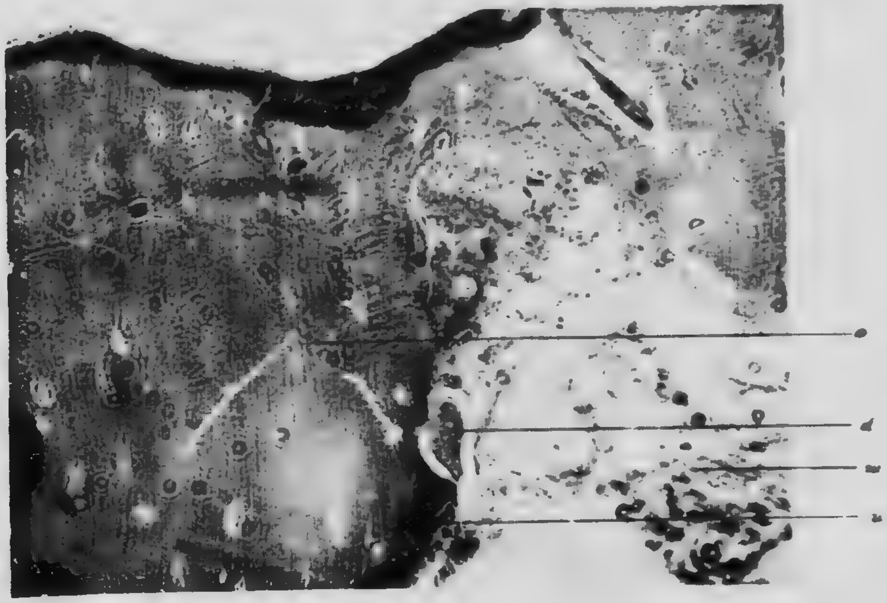
SECTIONS [illegible]

m. [illegible]

n. [illegible]

o. [illegible]

p. [illegible]



bone was much more marked than the deposition; in fact, the latter was altogether absent. In that case, old age was not the factor which produced the anatomical result so different from that hitherto associated with otosclerosis. Probably the very low state of health of the patient was the determining factor.

Returning to the description of the case under consideration, there only remains to be made a brief reference to the membranous structures. As has already been stated, the preparation had lain so long in weak spirit before it reached me that the membranous structures of the organ of Corti and ductus cochlearis had undergone disintegration to such a degree that no possible inference could be drawn as to whether or not any pathological change had occurred in these structures. With regard to the ganglion spirale, it may be stated that it did not show any changes which could be considered to be pathological. It has been previously pointed out, however, that this is merely negative evidence, and it is quite possible that pathological changes may have been present, but are not demonstrable by the present methods of preparation.

CHAPTER V

GENERAL CONSIDERATIONS ON THE PATHOLOGICAL CHANGES

In the first place it will be observed that in the four cases reported, various stages in the evolution of the disease as regards time are represented. Thus, the symptoms had been in existence for one year, three years, twenty-five years, and sixty years respectively. As regards the degree of deafness also, the symptoms varied from comparatively slight dulness to deafness of an extreme degree.

The ages at the time of onset were respectively 32, 48, between 25 and 30, and 20. In respect to sex, it is to be noted that all were female. Two were married and two unmarried.

With regard to the state of health at the time of onset of the symptoms, in two cases (III and IV) the health, so far as the patients could remember, was very good, and they could not attribute their deafness to any special cause. In Case I the patient felt quite well when the dulness of hearing began, but it is to be observed that she showed signs of phthisis a year later. In Case II the patient had been suffering from phthisis for two years previous to the first sign of deafness.

Inherited tendency to deafness was not known in the families of Cases I and II, but it should be stated that neither of these patients knew much about their relations. In Case III the grandfather was known to be deaf, but this may have been due to old age. An aunt on the paternal side was deaf, and this was not due to old age; but beyond that fact no more information could be gathered, so that the aunt may or may not have been deaf from otosclerosis. In Case IV non-suppurative deafness was a very marked feature of the family tree, and had been well recognised by the family for several generations.

Tinnitus was constantly present and was very distressing in Case III. It was also constantly present in Cases II and IV, but in neither of these was it distressing. In Case I it was hardly present at all, being only noticed on rare occasions, and even then only for a few moments.

Paracusis was definitely observed in Case II, but was not observed in Cases I and III. No reference was made to its occurrence in Case IV, so far as is ascertainable from the patient's friends.

Hearing was comparatively slightly affected in Cases I and II, but in Cases III and IV the deafness was extreme though not absolute.

Vertigo was not present in any of the cas.

The above synopsis of the clinical facts may be correlated with the corresponding synopsis of the facts revealed in pathological examination, which will now be given.

As regards the middle ear, no suppurative disease was present in any of the cases, but in Cases II and III there was a scar in the membrane of the ear of one side; but apparently this did not affect the hearing.

The mucous membrane of the tympanum was normal in all four cases, except Case II, and the Eustachian tubes were also unaffected in all, so far as post-mortem examination could show. Microscopic examination showed that there was no inflammatory activity in the mucous membrane over the promontory or elsewhere. In Case IV, however, there was some evidence of calcareous deposits, associated with a slight degree of fibrous thickening in the muco-periosteum over the diseased area of bone in the footplate of the stapes and pelvis of the oval window.

In all the cases the stapedius muscle and the tensor tympani were perfectly healthy in appearance. The striation was normal and there was no sign of fatty or other degeneration.

Symmetry.—Changes in the bone were present in all four cases, and in the three in which both ears were examined, the change was symmetrical, it might almost be said, to the minutest detail. In the fourth case only one ear was sent for examination, but clinically both ears were similarly affected, and it is, therefore, very probable that the symmetry was present in it also. In Case I not only was the symmetry of the bony lesion remarkable, but the actual phases of the osseous changes within the diseased area were, so to speak, remarkably symmetrical. Thus the portion of the diseased area in which absorption rather than deposition of bone was most marked, occupied the same anatomical position on each side. Again, in Case III the very fine bridge of bone passing from the footplate of the stapes to the wall of the oval window occupied exactly the same relative position in either temporal bone. In Case II the curious type of bony change, so different from that hitherto associated with otosclerosis, was symmetrical both in the character of the change and in the area affected. Finally, in regard to the symmetry of the lesion in all three cases, it may be pointed out that only one focus of diseased bone was found on each side.

In making these remarks upon the symmetry of the bony lesion, I am well aware of the fact that cases have been recorded, such as that of Manasse,[1] in which the lesion was not bilateral. Nevertheless, a study of the cases recorded by many observers leads one irresistibly to the conclusion that symmetry is a marked feature of the disease. The well-recognised clinical facts, of course, fully support that view.

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[1] Manasse, *op. cit.*

Another feature noticeable in all the four cases is the limitation of the area of diseased bone. This was particularly characteristic in Cases I, II, and III, and even in Case IV the anterior border of the diseased bone corresponded almost exactly with those of the first three cases. Indeed, we may say that so far as the anterior, external, and internal boundaries of the lesion are concerned, these are practically the same in all four cases. This is very remarkable when it is remembered that the duration of the disease in the four cases varied from one to sixty years.

As regards the posterior limit of the diseased area, Case III differed only from Cases I and II by a minute fraction of a millimetre, this minute portion consisting only of a bridge of bone replacing the annular ligament at one point, and causing ankylosis. In Case IV the posterior margin of the lesion was situated much further back, in the region behind the oval window.

In contrast with the remarkable similarity in the area affected in all four cases is the difference in the staining reaction. In Case I, with deafness of three years' duration, the portion of the diseased ar[illegible]ich absorption of bone was taking place, stained [illegible]ly, while that in which the deposition of bone was occurring stained very deeply. In Case II, with deafness of only one year's duration, and in which only absorption of bone was taking place, and no deposition of it, the whole area stained faintly. In Case III, with deafness of twenty-five years' duration, there was no evidence either of active deposition or absorption of bone, and the staining of the diseased area was almost identical with that of the surrounding normal bone. In Case IV, with deafness of sixty years' duration, the staining over all the anterior portion of the diseased area was almost identical with that of the surrounding normal bone, but in the extreme posterior portion of the diseased area, the staining was deep in the portions in which deposition of bone was occurring and faint in those in which absorption was taking place.

From these facts, we may infer that, in at least many cases of otosclerosis, the limitation of the diseased focus of bone is decided at a very early period of the disease, particularly so far as the anterior margin is concerned. In other words, the line of demarcation does not indicate an advancing edge, as some pathologists seem to think, but rather a limit within which certain changes have taken or are taking place, but beyond which these changes are not likely to occur, so far as that particular focus is concerned. In making this statement the writer is well aware that a number of cases have been described by other investigators in which the area of diseased bone is much larger than in any of the four cases recorded in the preceding pages. In such cases of more extensive areas of diseased bone, the explanation of their occurrence is to be found more probably in the coalescing of several foci in different parts of the capsule of the cochlea, rather than in the gradual extension of one focus.

The existence of a line of demarcation does not necessarily, or indeed frequently, indicate that there is present any special activity in the cellular changes in the bone. This is a very important matter, and the failure to recognise the fact has, I venture to think, led some writers into error in regard to the process by which the old bone is absorbed. Thus, Manasse, and following him, Panse, lay great importance on the fact that osteoclasts are not found in the region of the line of demarcation; and that therefore the absorption of the old bone is not carried out by osteoclasts as in normal bone.

Now, to judge from my own cases, I am inclined to think that this is an error, but a very excusable one. In the very great majority of cases of otosclerosis hitherto examined, the disease had been in progress for a long period of time, and in these cases osteoclasts are not found in the neighbourhood of the line of demarcation. Such a case, for example, is Case III, and the same applies to the anterior line of demarcation in Case IV. From an examination of a very early case such as Case I, however, the process of absorption of bone may be studied satisfactorily. It is particularly valuable in this respect, because within the focus of diseased bone one portion shows how absorption takes place by means of osteoclasts, while in the other portions the bone is being deposited and not absorbed, and osteoclasts are not found even along the line of demarcation.

Case IV is also instructive in regard to the changes which occur in the bone. There are two distinct areas which border on one another. The first is that which extends from the line of demarcation in front of the oval window to the other line of demarcation which lies very close behind the posterior margin of the oval window, and thus includes the whole of the footplate of the stapes. This is an old focus, and there is no more cellular activity in the bony tissue than in the normal portion of the capsule of the labyrinth. Osteoclasts, therefore, are not found. The second area of diseased bone lies behind the first, and is clearly seen to be in a state of cellular activity, and the process of absorption is going on so rapidly that the bone is becoming highly rarefied. Consequently osteoclasts are found in this area.

Thus, there are three types of change in the bone in otosclerosis. First, by far the most common type, if one is to judge by the total results of all investigators of the subject hitherto, is that in which absorption is followed by the deposition of bone, the new-formed bone being in excess of that absorbed. Cases I and III and the anterior portion of the diseased area in Case IV are of this type. Second, a type in which the absorption of bone is followed by deposition, but the new-formed bone falls short in amount of that which is absorbed. This is seen in the posterior portion of the diseased area in Case IV. Third, a type in which absorption occurs without any subsequent deposition of bone. Case II is an example of this type.

So far as the writer is aware, the second and third types have not previously been described.

A careful examination of Cases I and II throws grave doubts upon or perhaps even actually disproves Manasse's view that in otosclerosis the deposition of new bone is the primary change in the disease, and absorption of the old bone a secondary change, depending upon the pressure exercised by the newly deposited mass. In Case II there is no evidence of deposit of bone at all, but only of absorption. In Case I the most recently affected area is that in which there is no evidence of the deposition of bone, so that in this case also absorption cannot have been caused by the pressure of newly deposited material. In this respect my own investigations agree with those of Siebenmann.

In regard to the question as to whether the bony changes in otosclerosis are to be classed as inflammatory or not, opinions among those who have studied the problem are sharply divided. Siebenmann and Brühl hold the view that the process is not inflammatory, the latter referring to it as "formative." Manasse, Panse, and others look upon the bony change as being the result of chronic inflammation.

The result of my own investigations is entirely in favour of the view that the change is not associated with chronic inflammation. In none of my preparations was there any evidence of round-cell infiltration, and the sharp line of demarcation is a feature hardly consonant with a pathologist's conception of chronic inflammation.

Habermann's view of the pathological process is that the change in the bone is the result of chronic inflammation in the periosteum of the tympanic cavity. It is of interest to note that at one time Politzer held this view, but he has since given it up and now holds that the characteristic change occurs in the body capsule of the labyrinth.[1]

When otosclerosis was first described as a change in the bony capsule of the labyrinth, it was supposed that ankylosis of the stapes was an essential sequence upon the pathological change. Even now, Brühl would consider that the cases in which this occurs should be separated from the others, and should alone be included under the name of otosclerosis. It is, however, difficult to see why the term should be limited in this way, unless it be for purely clinical reasons. The fact that the most common position at which the process begins is the region immediately in front of the oval window does not indicate any fundamental difference in regard to the pathological changes. These are the same, no matter what part of the capsule of the labyrinth is affected. The present writer's explanation of the fact that the region referred to is particularly liable to undergo these changes, is given below, and depends in part also upon the evolution of the organ (see p. 57). The same explanation also accounts for the fact that the capsule of the cochlea is much more frequently affected than the portion which surrounds the rest of the vestibule and the semicircular canals.

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[1] Since this was written, Fraser * has demonstrated a very interesting case of otosclerotic foci occurring in the capsule of the labyrinth in a case of suppurative middle ear. This case confirms by direct pathological observation what I have deduced from clinical evidence (see section on Heredity), that suppurative middle ear disease may be the immediate factor in otosclerosis. It does not, however, prove that otosclerosis is always or even frequently the result of middle ear infection.

* (*Proc. Roy. Soc. Med.*, May 19th, 1916.)

Brühl's explanation of the fact that the most common position for the bony change is in front of and above the oval window is ingenious. It is, that the play of the tendon of the tensor tympani in close proximity to this region determines, in many cases, the initial focus of the disease in the bone. I think it quite probable that this may be a factor, but there is another which I think as important. This is the stress which is imposed upon the walls of the oval window by the movements of the stapes. It must be remembered that when the stapes moves, strain is put upon the fibres of the annular ligament, and, through them, upon the walls of the oval window. Now, the lines of stress do not cease at the walls of the oval window. Movement ceases there, but the lines of stress are radiated through the surrounding bone all the same, and the living cells in the bone will respond to this by an increased activity in some way. In those who have a predisposition to otosclerosis the result of this increased activity will be the establishment of the disease first at that region.

Further, this explanation accounts for the fact, so well known clinically, that a considerable number of cases of otosclerosis are associated with chronic nasal or naso-pharyngeal catarrhs. Indeed, many really are mixed, and the mistake is made in diagnosing them as cases of purely nasal or Eustachian origin. In the light of the explanation given above, it is not difficult to follow the sequence of events. The chronic catarrh in the nose or naso-pharynx leads to more or less Eustachian obstruction, with resulting indrawing of the tympanic membrane and chain of ossicles. Owing to the inward pressure, the ligamentum annulare is in a constant state of hypertension, and transmits the stress to the walls of the oval window and the bone in the neighbourhood, as described above, with the result that in those in whom there is any tendency to otosclerosis, the bony change is prone to manifest itself in that region. Even in these cases, however, the fundamental principle that the pathological process in otosclerosis depends upon the evolution of the organ of hearing, still holds. For we find that the bony change tends to arise in front of the oval window rather than behind it, and in its later extension affects the bony capsule of the cochlea, while with the rarest exceptions, the bony capsule of the semicircular canals remains unaffected.

It is interesting to consider the period of time in the course of the disease at which ankylosis occurs in the stapedio-vestibular articulation; but it must be admitted that the conclusions arrived at are for the most part speculative. In Cases I and II, in which the disease had been in existence for one and three years respectively, there was no sign of the involvement of the articulation in the bony change. In Case III, in which the disease had lasted twenty-five years, bony ankylosis had actually occurred. But in this case it must be pointed out that the trabecula of bone uniting the stapes to the wall of the oval window was extremely thin, occupying a depth of about 40 or 50 μ, and was only present in a very limited region at the anterior extremity of the oval window. Further, the bony change extended only a very short distance into the footplate of the stapes. All these facts, however, are no proof that ankylosis had occurred recently. Indeed, the fact that the otosclerotic bone in this region did not stain any more deeply than the normal bone shows that the change cannot have been of very recent date. In Case IV, in which the disease had been present for over sixty years, the footplate of the stapes was united by bone in all its circumference to the walls of the oval window.

We may conjecture, therefore, that ankylosis is not usually an early feature of the disease, and may in some cases be a comparatively late one, and perhaps may never occur at all. But further investigation on this subject is necessary before we can speak with any degree of certainty.

Leaving now the consideration of the changes in the bony capsule, which are so characteristic of otosclerosis, the question arises as to the part which the nerve structures play in the disease.

It will be seen that in three of the cases recorded in this paper, the sound-perceiving apparatus showed no sign of pathological change, and in the fourth case no decision on this point could be given. The same normal condition of the sound-perceiving apparatus has been described by other investigators.

On the other hand, it is unquestionable that some cases of otosclerosis show marked degenerative or atrophic changes in the nerve structures of the labyrinth. Manasse has described a most instructive case in which the bony changes characteristic of otosclerosis were associated with marked pathological changes in the nerve structures of the membranous labyrinth in one ear, while in the other ear the bony capsule of the labyrinth was normal, and the nerve structures were markedly affected.

Again, it appears, judging from clinical evidence, that the sound-perceiving structures tend ultimately to become involved as time goes on.

Finally, it must be remembered that even when nerve structures present a perfectly normal appearance under the microscope, we are by no means justified in stating that they are really normal in the physiological sense of the term. As proof of this may be cited the fact that in some cases of ordinary neuralgia even of long duration, the microscopic examination may reveal no pathological change in the nerve structures, and yet it is inconceivable that there really is no change. Also, as in Case III, tinnitus lasting for twenty-five years produced no discoverable change, yet surely *some* change must have been present in the nerve-structure. In such a case we can only say that there must be a pathological change of some kind, but our present methods of examination fail to reveal it.

How then are we to look upon otosclerosis from a pathological and biological point of view?

As previously stated, Brühl considers that the cases in which there are bony changes in the capsule of the labyrinth associated with ankylosis of the stapes are different from the others, and that the term otosclerosis should be restricted to these. Manasse, on the other hand, maintains that such a nomenclature is artificial, and that the term otosclerosis should be abolished. His reason for this point of view is that neither clinically nor pathologically is it possible to separate the groups.

Manasse's view would probably commend itself to clinicians better than Brühl's, because it is undoubtedly the case that very frequently the clinical symptoms and signs indicate both a bony change in the capsule with ankylosis of the stapes, and, in addition, a change in the sound-perceiving structures. The difficulty in accepting Manasse's view is that we have not hitherto known of any general biological or pathological principle which would account for this frequent association of the two types of cases, viz. those in which the bony capsule of the labyrinth alone is involved, and those in which the sound-perceiving apparatus is coincidently affected.

Now, if the whole question be considered from the point of view of the evolution of all the structures of the organ of hearing, the problem becomes clearer.

The organ of hearing must be considered as a unit composed of many different anatomical parts, each of these parts having been evolved in intimate correlation with one another. For example, the formation of a drum membrane and chain of ossicles is associated with the formation of an oval window, allowing the vibrations to be carried in to the nerve terminations. The formation of an oval window is associated with the formation of a round window, allowing the movements of an incompressible fluid to take place. The increasing development of the neuro-epithelium is associated with the absorption of bone or cartilage to form ultimately the cochlea, this change being necessarily accompanied by activity in the bone and cartilage cells surrounding the corresponding neuro-epithelium. And so on with all the structures concerned, including all the neurones associated with the function of hearing (see p. 57)

The whole organ of hearing from the cerebral cortex to the auricle must therefore, from the point of view of its evolution, be considered a biological and physiological unit ; and the anatomical portions, though so different from one another in tissue and structure, are intimately correlated in their evolution.

In the fertilised ovum, therefore, according to the view presented in the previous section of this paper, we may consider that there are inherited, not only potentialities for the development of the various tissues or structures of which the whole organ of hearing is composed, but also a potentiality presiding, as it were, over the correlation of these tissues and structures to one another.

Now, since, in accordance with biological principles, these potentialities cannot be possessed in exactly equal degree in different individuals, it is not difficult to see why the organ of hearing should, in some individuals, fail to withstand injurious influences, while in other individuals, in whom these potentialities are more strongly marked, it will be able to resist such influences.

Further, since these potentialities are inherited, it follows that the pathological changes which are included under the term otosclerosis will tend, on the whole, to show evidence of inheritance, just as is frequently the case in deaf-mutism.

It will naturally be asked, however, if the explanation of the nature of otosclerosis just given be assumed, why should the change in the bone be so prominent a feature in the disease in comparison with other changes, as for example in the nervous system, the mucous lining of the tympanum, etc.? There are very good reasons for this. In the first place, aural pathologists have in the great majority of cases made a special point of looking for changes in the bone, whereas I know of none (myself included) in which careful macroscopic examinations have been made of the cerebral cortex in the temporo-sphenoidal lobe, or in the auditory nuclei of the medulla, or of the nerve-fibres and glandular structures of the middle ear, or of the similar structures in the external meatus and tympanic membrane. This point is particularly important, because we know clinically that there is, in at least the majority of cases, a marked diminution in the sensitiveness of the tympanic membrane and external meatus, as well as a diminished secretion of wax. Changes, therefore, must be present in these structures, but pathologists have never looked for them. Again, it is by no means unreasonable to suppose that the prominent symptom of tinnitus may really be due to pathological changes in the neurones of the cerebral cortex and not to changes in the labyrinth at all. If such a supposition proves ultimately to be correct, it would explain why mental exhaustion, worry, etc., have such a serious effect upon the course of the disease, and also why destruction of the cochlea and section of the auditory nerve do not always cure the tinnitus.

But there is another reason why the bony change should appear so prominent in the pathological as distinguished from the clinical picture. In all the tissues with which we are at present concerned, except bone, cellular changes in the sense of the death or disintegration or disappearances of cells which are substituted by others, is a comparatively rare phenomenon unless inflammatory activity is present, and this is not the case in otosclerosis. Thus, the neurone is never replaced by another neurone; and except for accidental damage, it lasts throughout the life of the individual. So far as is known, glandular and epithelial and connective tissue cells have a very long life. But with bone the case is different. In this tissue the disappearance of old cells followed by the arrival of new cells is the normal condition of the tissue. If, then, a very slow degenerative, as distinguished from an inflammatory, change takes place in the various tissues comprising the organ of hearing, it is not surprising that such changes will appear more prominent to the eye in the case of bony than other tissues. The meaning might be made more clear by expressing it in another way. Thus, let it be supposed that constant but slow reproductive change is going on in the cells of a normal portion of epithelial tissue, so that at the end of, say, two years all the original cells have disappeared and been replaced by new ones. And let it be again supposed that a similar piece of epithelial tissue is subjected to a degenerative pathological process, so that all the original cells were replaced by new ones. Now, microscopic sections made through these two tissues respectively would not show any very noticeable difference, because at any given time the cellular changes are really very slight. On the other hand, if in a tissue such as normal bone all the old cells of the tissue were by the end of, say, a month replaced by new ones, and if in another piece of bone all these old cells were replaced within a fortnight, then the difference in the staining and other properties of the two pieces of bone respectively would be quite striking to the eye.

The lesson to be learnt from these considerations is that, as pathologists, our attention has been concentrated too narrowly upon the bony capsule of the labyrinth, to the exclusion of other parts of the organ of hearing. In the future when an opportunity arises of examining a case of otosclerosis after death, the investigator should make a careful examination of the temporo-sphenoidal lobe of the cerebral cortex, of the auditory nuclei in the medulla, and of the nerves and glandular structures in the middle ear, the tympanic membrane and external auditory meatus. The superior cervical ganglion of the sympathetic, as also the ganglia of the pneumogastric, the glossopharyngeal, the facial nerve, and the fifth nerve should also be investigated. In this way it may be found possible to explain certain well-recognised clinical facts, such as the occurrence of tinnitus, the peculiarly serious effect which mental exhaustion has on the patient, both in regard to the tinnitus and the deafness, the diminution in sensitiveness of the tympanic membrane, and the diminished secretion of wax. None of these clinical facts is explained by the changes in the bony and membranous labyrinth.

The same objection applies to the theory recently put forward, that otosclerosis is due to activity in the islands of cartilage in the bony wall of the labyrinth, resulting in the deposit of new bone. It may be true, of course, that these islands of cartilage become active when otosclerosis occurs, and, indeed, such a condition is referred to in Case II. But this is only one of the manifestations of the disease, and not the essential causative factor. Any satisfactory theory of otosclerosis must explain the occurrence of both the clinical and pathological phenomena of the disease, and all those theories which attribute these phenomena to anatomical changes limited to the bony wall of the labyrinth fail in this respect.

In viewing the subject of otosclerosis in this way, it is now possible to understand the occurrence of the peculiar signs and symptoms of the disease, and why these vary in different cases.

First.—The inheritance of the condition. This has been investigated in detail in the section under that heading, and need not be discussed further at present. The fact that otosclerosis frequently occurs without evidence of inheritance has also been explained in the section mentioned.

Second.—The absence of any signs of inflammatory activity. Otosclerosis is a degenerative process, and therefore may occur without any sign of inflammation.

Third.—The diminished secretion of wax, so characteristic of the disease, is due to the fact that the organ of hearing, being an evolutionary unit, is failing more or less in all its parts.

Fourth.—The loss of sensitiveness of the meatus and tympanic membrane is explained on the same grounds as the last; and if opportunity offered it would probably be found that the mucous membrane of the tympanum participates in this loss of sensitiveness.

Fifth.—According to some writers, one of the symptoms of otosclerosis is an abnormal patency of the Eustachian tubes. It is difficult to prove that the Eustachian tube is wider in the subjects of otosclerosis than in normal individuals, because, as Fraser aptly remarks, catheterisation of normal individuals must be a rare occurrence, to say the least; and we have, therefore, no accurate standard by which to judge. Nevertheless, on the present writer's theory of otosclerosis, undue patency of the tubes might almost be expected, because of the diminished blood supply which is associated with atrophic and degenerative processes.

In accordance with this view, the involvement of the nerve structures in the cochlea, which is frequently present in many cases of otosclerosis, is not to be looked upon as a result of changes in the bone, but as a part of the degenerative change in the organ of hearing as a whole. It probably begins in otosclerosis long before we are able to demonstrate it by microscopical methods, and may perhaps account for the tinnitus which is so common a feature of the disease. But this symptom might also be accounted for by changes in the cerebral cortex.

This theory accounts for the absence of pathological and clinical disturbances in the vestibule and semicircular canals. It is a degenerative process in the organ of hearing, not in the organs of equilibration.

From the foregoing pages we may gather indications for the treatment of otosclerosis, but the details of treatment are discussed in the appropriate section. At present the following points may be emphasised:

1. By no process can we change the innate tendencies of the cells and structures of the organ of hearing. They are characteristics of the individual which he must carry with him through life.

2. Since toxic and other constitutional conditions, as probably also local ones, tend to render these potentialities active, we can, within limits, use means to keep them latent, or restore them to a latent condition—if they have become active.

3. There is no constant single constitutional or local exciting cause in otosclerosis, but various local or consti-

tutional factors may be the exciting cause in different cases. Hence the search for a specific line of treatment is in vain. Each case must be treated individually.

In conclusion, a few words may be said in regard to the nomenclature of the disease. "Otosclerosis" is quite a harmless name if we are careful not to attach any significance to its derivation; and no one is likely to be led far astray by a definition so obviously remote from anatomical or pathological precision. Like the terms "cancer" and "syphilis," it has the great advantage of not pretending to indicate a knowledge of the disease which we do not really possess.

On the other hand, all names which include the term "ostitis" are undesirable, because in the first place they suggest the idea that the pathological change is limited to the bone, and in the second place they indicate inflammatory activity. Both of these conceptions are incorrect. The term "osteoporosis" is open to one of the above objections, but it is good in so far as it does not suggest the presence of inflammation. The name "Progressive Deafness" is too vague, and, moreover, might include cases of double suppurative middle ear disease, as well as other conditions certainly not meant to be included.

A name which the writer ventures to suggest as being more satisfactory than Otosclerosis is, "Idiopathic Degenerative Deafness." The term "idiopathic" is used in the sense that the disease is liable to occur in certain individuals and can occur in those individuals only. The word "degenerative" indicates that the disease is of the nature of a retrograde process and not of an inflammatory one. The term has the advantage of not assuming that the disease is confined entirely to any one structure or tissue of the organ of hearing.

Investigation in Respect to Some of the Inorganic Constituents of the Blood in Otosclerosis

On the suggestion of Major E. B. Waggett, I considered it desirable to make an investigation of the blood in cases of otosclerosis in respect to the occurrence of some of the inorganic constituents, so that a comparison with normal blood might be made.

The analysis was undertaken by Dr. W. Hunter Duncan, Pathologist and Bio-chemist to the Prince of Wales's General Hospital, and the results of his investigation are given here.

In all, seventy-one specimens of blood were analysed, and of these forty-four were of normal blood and twenty-seven were of the blood from cases of otosclerosis.

Normal Blood.—One hundred parts of normal blood-plasma yield :

Total solids (ash)	0·8554%
Chlorine	0·3644%
Sulphur trioxide (SO_3)	0·0115%
Calcium (as phosphate)	0·0313%
Magnesium (as phosphate)	0·0225%

Otosclerosis.—In *otosclerosis*, the amount of *chlorine* was variable in the twenty-seven cases, but the average was 0·3641%, being thus 0·0003% below that of normal blood.

The *sulphur trioxide* (SO_3) in the cases of otosclerosis showed a maximum of 0·0114% and a minimum of 0·0112%. Even at the highest, therefore, the amount is 0·0001% below the average of normal blood.

The *calcium* (as phosphate) content in otosclerosis was found to average 0·1002% ; this being an increase of 0·0689%. It would appear, therefore, that in otosclerosis there is an increase of calcium to the extent of fully three times that of normal blood.

The *magnesium* (as phosphate) content in the blood from the cases of otosclerosis was 0·0301%, showing an increase of 0·0076%. That is an increase of about one-third of that of normal blood.

In regard to the occurrence of *sodium* and *potassium*, an attempt was made to ascertain their content in the blood.

They varied, however, within such wide limits that no satisfactory conclusions could be drawn from the percentages found to be present.

As result of this analysis, it may be said in general that in so far as the figures relating to the sulphur trioxide and the chlorine are concerned, the difference between normal blood and that from the subjects of otosclerosis is so small that it may be considered negligible.

In the case of the calcium and magnesium content, however, the subject requires further consideration. The presence of calcium in the form of phosphate to an extent three times greater than in normal blood, would appear to indicate a metabolic change in the body of some kind different from those of healthy individuals. Unfortunately, however, we still remain in the dark as to what the nature of that change may be.

It may be pointed out, in order to prevent any misconception, that the greatly increased calcium and magnesium content in the blood can have no quantitative relationship to the change in the bony capsule of the labyrinth in otosclerosis. That is to say, the absorption of lime and magnesium salts which takes place in the region of the diseased bone in the capsule of the labyrinth is far too small to account for the change in the chemical constitution of the blood, and indeed is probably so small as to be impossible of detection by chemical means.

There may conceivably, however, be a slight change throughout the whole skeletal structures of the body in otosclerosis, sufficient to produce the change in the percentage of calcium and magnesium elements in blood. But such an explanation is very difficult of acceptance, because clinical experience would surely give evidence of such a change.

At present, however, it is idle to speculate further upon the significance of the increased calcium content in the blood shown by Dr. Duncan's analysis. The results of the analysis are given here rather as a record for corroboration by further investigation and as an indication for future lines of research.

CHAPTER VI

NOTES ON THE TREATMENT AND PROGNOSIS OF OTOSCLEROSIS

In the following pages some methods of treatment, devised both by other aurists and by myself, are considered.

One of the methods of treatment which has gained considerable notoriety, both among the medical and lay public, is that devised by Heath; and it may, therefore, be worth while to give the results of my own experience with it. It consists essentially of the application of blistering agents to the tympanic membrane. The method is not based, so far as the present pathological investigations show, upon any definitely known facts of the morbid anatomy of the disease; and it must not be judged on these lines at all. A method of treatment of a diseased condition might conceivably be quite valuable, even though it may not be based on known pathological facts. Judgment must in such cases be made only as the result of trial of the method, and I therefore carried out the treatment advocated by Heath in a number of cases, which fulfilled the clinical conditions which he maintains indicate the application of the method he devised.

Of the patients selected for trial, only twelve submitted themselves to the treatment for two months, and these are taken as the basis upon which judgment as to the value of the treatment is to be made. The remainder, either on account of the disagreeable nature of the treatment, or from lack of faith in it, or from some other cause, gave up attendance before the two months were complete. Of the twelve cases which were treated for two months, none were treated for more than three.

Before treatment was begun, measurements were taken of the patients' hearing power, with the watch, the whispered voice, the conversation voice, the high notes and low notes. The condition of the bone conduction was also noted. Measurements were again taken at intervals during treatment, and also after treatment at periods varying from a fortnight to six or eight months.

There is no need to give details of each case, for the ultimate result was that in not a single case was there any improvement in the hearing at any time. It is of great importance, however, to observe one fact in relation to several of these cases, because it explains why this method of treatment has had a vogue among the lay public. This fact is, that after the discharge and reaction which result from the blistering cease, as they do after the treatment is stopped, a number of patients are convinced that they hear better as a result of the treatment. When the hearing tests are applied, however, it is found that there is no actual improvement, the hearing being the same in every respect as before. The explanation of these apparently inconsistent observations was, however, not difficult to ascertain. As stated above, measurements were taken of the hearing power *before*, *during*, and *after* treatment. On investigation of the figures it was found that the hearing power began to diminish as soon as the inflammatory reaction to the blistering process set in, and this reaction was maintained during the whole treatment, the hearing power being kept at a low level for the period of the two months. At the end of this time the patient has forgotten what his hearing was like before treatment, and consequently when reaction subsides and the hearing improves up to its former condition, he is sometimes convinced that his hearing has improved. But on applying the tests the illusion is dispelled.

Before dismissing this subject, it must in fairness be said that the hearing is very rarely adversely affected by this treatment. It almost always returns to exactly the same condition as before treatment. In one case, however, that [illegible] hospital nurse, the result was unfortunate. In [illegible] case perforation occurred in the membrane as a result of the treatment, and since then she suffers from periodic short attacks of otorrhea, and this still goes on, although it is a year and a half since the treatment was given up. The hearing in the ear which was treated has been distinctly worse, and the tinnitus is certainly no better.

In another case, not of my own, but of which I heard indirectly, the result was still more serious. Acute middle ear inflammation occurred, followed by mastoiditis, which required operation. The method of treatment is, therefore, not free from danger.

Of the various methods of treatment by the so-called re-education of hearing, it is only necessary to refer to one of the better-known ones, the Zund-Burguet method. This is only a modification of the principle originally suggested by Urbantschitsch. I have not been able to carry out the method myself, but three of my patients who had had their hearing tested previously, underwent a course of treatment by the Zund-Burguet method. After the treatment, no improvement occurred in two of the cases, and in the third, the hearing distance for the watch had risen from 6 ins. to 7 ins., a degree of improvement which may be considered negligible, especially as the patient was unaware of any difference in the hearing and the improvement soon disappeared. Tinnitus was present in two of the cases, and in these no benefit occurred as a result of the treatment. Thus there is no ground for the belief that this recent application of the principle of re-education of the hearing offers any advantage over the others which preceded it.

The psychology of individuals who suffer from dulness of hearing is an interesting study; and the vogue among the public, which these methods have had, illustrates the fact when large numbers of individuals are taken. The great majority of people can be persuaded, and usually very easily persuaded, that they hear better as the result of treatment of any kind directed to the ear. Only to the minority, possessed of critical judgment, does it occur to try by simple but genuine tests whether their hearing has improved or not. It is in consequence of this that practically any method of treatment can, for a time, acquire popularity if it be sufficiently advertised and talked about. This fact has been brought forcibly home to me in the course of the present investigation in regard to the following method devised by myself.

At the outset of the investigation, and before I had reached the conception of otosclerosis which I now hold, it occurred to me that, although sudden and practically instantaneous attempts to mobilise the ossicles as, for example, by the catheter or inflation, had failed other methods might be successful. Very slight pressure brought to bear upon the chain of the ossicles over a considerable period of time might effect what a sudden jerk was unable to do. I therefore made use of the expansile qualities of compressed cotton-wool in the following manner. Little pellets of cotton wool were compressed tightly and then inserted, one by one, into the meatus, care being taken that the innermost were in actual contact with the membrane. When the meatus was almost full of these, an india-rubber cork was inserted tightly into it so that, as the cotton-wool expanded, the tympanic membrane along with the chain of ossicles would very gradually be pressed inwards. The cork was kept in for a period of from one to three or four hours, and was then removed along with the cotton-wool. This procedure was undertaken twice weekly for two or three weeks. It was employed in six cases. The results of the treatment, as gathered from the patients' statements on the one hand and from actual testing on the other, were very interesting. Of the six patients, four stated that the hearing was distinctly better, and these statements were made voluntarily, without any attempt at suggestion. The remaining two patients said there was no improvement. When, however, examination of the hearing was made, it was found that no change had taken place in any of them. In respect to the tinnitus there is, of course, no objective test, but of the six patients, three suffered from fairly severe tinnitus, and of these three two were of opinion that it was unchanged as a result of the treatment, while the third was very emphatic in maintaining that it had improved very much.

This is, therefore, another good example of the difficulty of judging from patients' statements of the improvement in the hearing as a result of treatment. Otologists are right in demanding that no reliance can be placed on any claims as to improvement in the hearing as the result of treatment by this or that method unless definite measurements can be given. Indeed, it would be better in such an important matter as this, that any aurist who wishes to establish a claim as to a new method, should first submit the patients to inspection by unprejudiced minds, and then, after treatment, again have them examined by those who had made the preliminary inspection. Unfortunately, aurists have to suffer opprobrium on account of the no doubt well-meant, but mistaken enthusiasm of a few who imagine they have discovered an improved method of treatment. In any case otologists should be very critical in their attitude towards such claims, as they have caused much disappointment and suffering to the victims of otosclerosis, by inspiring them with hopes which have not been realised.

General Considerations

If the reader has considered the portion of this monograph which deals with the pathogenesis of otosclerosis, and also the chapter dealing with the hereditary tendency and environment, he will readily understand why very great importance is attached by the writer to the consideration of the general condition of the patient. This is really the basis on which not only treatment, but also, to a great extent, prognosis will depend.

In the portion dealing with inherited tendency I have attempted to view, with such critical judgment as I possess, the extent to which the onset of otosclerosis is determined by inherited tendency or potentiality on the one hand, and by environment on the other. I have shown further, that no sharp line can be drawn in the way of separating out one group from the other, or referring to one set of cases as being due to inheritance, and another not.

With regard to otosclerosis, as probably also with regard to many other pathological conditions, the human race may be looked upon as a series in a scale. At one end of the scale are found the individuals in whom the potentiality for developing otosclerosis is so very insignificant that it may be considered negligible; and no environment, whether acting locally or constitutionally, can stimulate it into activity and produce otosclerosis.

At the other end of the scale are those in whom the potentiality is exceedingly strong: so much so that in them practically no special stimulus or unfavourable environment is necessary to call otosclerosis into existence. In the latter the ordinary physiological processes in the body are sufficient for the purpose, transforming what is only a potentiality into an actuality.

It has further been shown that there is no single exciting cause of otosclerosis. In other words, from a given individual or personal potentiality, one or more of many exciting causes may call otosclerosis into existence in such an individual. Indeed, in the same individual the exciting cause of otosclerosis in one ear may be different from that which produces it in the other ear. An example of this is related in Case B1,[1] p. 34, in which acute middle ear inflammation caused otosclerosis in one ear, while some other cause not definitely proved, but certainly not acute middle ear inflammation or any other middle ear trouble, caused the disease in the other ear fifteen years later.

It is also of great interest to observe that in cases in which there is a strong inherited tendency to otosclerosis, the exciting factor may be quite different in different members of the family. This is shown in Cases C3, C7,[2] pp. 25, 27, and it is of further interest to note that it explains why, even where the inherited tendency is clearly present, yet the results of treatment are very different. One brother or sister of the family may react well to a given line of treatment, while another may not respond at all to that or any other treatment. The significance of these remarks, then, is obvious. There can be no routine treatment for otosclerosis, and the hope for any such is chimerical. Every case must be a study in itself, and if properly pursued the study is fascinating to the last degree. It must almost always be difficult, and the result of treatment, so far as improvement goes, is generally unsatisfactory; nevertheless if the study of each case is carried out thoughtfully and with tenacity, it is remarkable how often the course of the disease may be influenced in so far as preventing further progress is concerned; and in some cases even considerable improvement may be brought about. Further, if each case is taken individually the physician may be able to say when there is reasonable hope for improvement or arrest of the disease, and when there is not.

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[1] See chapter on Heredity.

[2] See chapter on Heredity.

The general considerations which the writer wishes to emphasise are several. The first of these is to take a rational and philosophical view of the disease, based upon a knowledge of pathological processes in general and of otosclerosis in particular, and upon the family and personal history and clinical symptoms. These must be made the subject of individual consideration.

The next point of importance is that the physician must disabuse his mind, on the one hand, of the idea that he has little or no control over the disease, and on the other hand of the expectation of achieving very striking results from the patient's point of view. The first involves unnecessary pessimism; and those who maintain the second are too uncritical and too enthusiastic. It is true, indeed, that in some cases quite noticeable improvement occurs in otosclerosis if the line of treatment suited to the particular case is faithfully carried out, but in the majority the improvement, if any, will be slight. On the other hand, the pessimist who looks upon all cases of otosclerosis as altogether outside his control may do great injury to his patients owing to his incorrect, though honestly held opinion.

Speaking in general terms, the truth is approximately as follows: Some cases will steadily become worse in spite of all or any treatment. In a large number, especially if seen in the very early stages, the course of the disease may be arrested or its advance may be greatly retarded. In a few some degree of improvement may be obtained, and in a very few the improvement may be considerable. In no case that I have ever seen was the hearing restored to the normal.

To repeat, there is no routine treatment for otosclerosis. The conditions of its occurrence are far too varied and far too subtle for any rule-of-thumb consideration. Every aurist, however skilful and patient, will certainly have many failures to record ; but, taken over large numbers, he will be most successful who possesses the requisite patience clinical insight, imagination, and judgment.

It is impossible to give a list of all the bodily conditions which may be the exciting cause of otosclerosis, and therefrom form the basis of treatment. But in the following pages indications of some of these will be shown with cases to illustrate them. Before doing this, however, it is desirable that some words should be said in regard to prognosis, and I will attempt not only to indicate the facts which should guide us in respect to prognosis, as has been frequently done before by others, but also, guided by a knowledge of the pathogenesis of the disease, to show why these facts affect the prognosis. In this way the study of the disease is rendered more interesting and it becomes rational rather than empiric.

Age of Onset.—It is well known to every aurist that prognosis is guided to a certain extent by the age of onset in a given case. Especially is this true when the age of onset is very early. Thus, although otosclerosis is rare under the age of thirteen or fourteen, yet, when it does make its appearance then, the outlook is extremely grave. I have never seen such a case appreciably helped by treatment ; and I venture to think that this is the experience of most aurists who are careful not to mistake Eustachian catarrh or middle ear catarrh for otosclerosis.

Now, if otosclerosis were an inflammatory condition, exactly the reverse of this would be expected. The recuperative powers are much greater in the young, and it would be expected that this would enable the sufferer to overcome the condition if put under proper environment, if it were the case that the disease were inflammatory in character.

It is of interest, in passing, to note that otosclerosis is not the only disease in which this peculiarity in regard to prognosis holds true. The same is found to be the case in malignant disease, in arterio-sclerosis, and in diabetes.

Heredity.—When otosclerosis makes its appearance in an individual in whose family there is an obvious hereditary tendency to the disease, the prognosis is on the whole rather unfavourable. This fact is well known to aurists. The explanation appears to me to be that the factor which is, for the most part, responsible is in general more likely to be innate in the cells of such an individual than in those who come of a normal family. In these cases, therefore, other unfavourable constitutional conditions or environment play a smaller part in the case. One would expect that such cases would, in general, respond less readily to treatment than those in which the disease arises for the most part as a result of some factor which can be definitely removed, and in which the innate tendency in the patient is not so pronounced.

Tinnitus.—The presence of tinnitus in a pronounced degree is well known to indicate a condition rather likely to progress rapidly. The reason for this is somewhat obscure, and the following suggestion is only tentative. It has been shown that otosclerosis is not a condition affecting only one portion of the organ of hearing. It is a degenerative process in which the nerve structures, walls of the labyrinth, and even the secretory apparatus of the external meatus, as well as the sensory nerves supplying the tympanic membrane, may all be involved. It has also been shown in the section on pathogenesis why this should be so. It may be true, as Manasse maintains, that the nerve apparatus of the cochlea and auditory nerve are really the primary structures to depart from the normal, and that changes in the bone and other structures are secondary; but this, it must be noted, does not mean that the secondary changes in the bone, if such they be, must be in exact proportion to the extent of the disease process in the nervous mechanism. A comparatively slight departure from the normal in the sound-perceiving apparatus may, by its effect on the correlated vasomotor nerves which regulate the blood-supply to the bone and other structures of the sound-transmitting apparatus, bring about very considerable changes in the latter. Now, when viewed from this aspect, it is not very difficult to see why the cases in which severe tinnitus is present are those which offer less hope of relief or arrest of progress. They are the cases in which the nerve structures are particularly involved, and, as is usually the case when disease of nerve structures are concerned, there is comparatively little hope of improvement. There are, however, a few exceptions to this rule, and amongst them the most noticeable are those in which the ear affection arises in women during a time of chlorosis. Such cases are often amongst the most satisfactory to treat, in spite of the fact that in many, tinnitus is a fairly marked symptom. The reason, of course, why the treatment is so comparatively satisfactory in these cases is that the chlorosis which is the exciting cause can be definitely and certainly removed, with the consequence that the aural malady ceases to progress and frequently improves considerably. Further, in these cases which improve, there is a fair probability that the bony change in the capsule of the labyrinth has not progressed very far and the stapes may not be ankylosed. Indeed, in the cases occurring during chlorosis, there is some hope of improvement even when an hereditary tendency towards the disease is present in the family.

Paracusis.—It is a remarkable fact that aurists hold very different views as to the significance of paracusis from the point of view of prognosis. The majority consider its presence a sinister omen, but Yearsley and some others are of the converse opinion, and view the condition as one indicating a comparatively satisfactory result if treatment be carried out. My own experience in the matter is that, while paracusis is of ominous significance so far as improvement by treatment is concerned, it is good to this extent, that such cases tend to progress rather slowly, and may even be arrested if treatment can be satisfactorily carried out. This statement is particularly true if paracusis is present without tinnitus.

Climate.—In speaking of climate in regard to its effect upon otosclerosis, it must be understood that the disease is considered in its pure form, unassociated with coincident naso-pharyngeal, Eustachian, or middle ear catarrh. When these are present, of course, a moist climate is notoriously bad. But in cases of otosclerosis uncomplicated in the manner indicated, I have never seen any reason to suspect that moisture in the air, *per se*, has a deleterious effect.

Severe cold is particularly bad for otosclerosis, and this applies to a cold dry climate as much as to a cold moist one.

Diet.—There is very good reason to suppose that the course of otosclerosis can be affected, to a certain extent, by diet. This statement is based upon the principle of the pathogenesis of the disease described under that section, and upon experience. On the supposition that the pathogenesis of the disease is as indicated, then it may well be understood that a diet which permits of considerable toxin formation and absorption during the process of digestion, will, on the whole, have a bad effect. A diet consisting largely of meat should, therefore, be avoided; and it is advisable to restrict the amount of animal food to the minimum that is compatible with perfect health. In otosclerosis the man who, as the phrase goes, "does himself well," most emphatically does himself ill.

Alcohol and tobacco, as is well known to all aurists, have a bad effect upon the disease, as also have strong coffee and tea, though perhaps in a lesser degree than the two former. Other conditions governing the prognosis are too well known to require more than enumeration. Such are, exhaustion, want of sleep, foul air, and excess of any kind.

There is one factor, however, that I have observed, but have not seen referred to elsewhere—injury, especially injury to the head. A case illustrating this is referred to on p. 42,[1] in which the injury was probably the immediate exciting cause of the otosclerosis, and I have met with several cases in which injury appeared to make the deafness and tinnitus very rapidly worse. This is not difficult to understand when we consider the shock to the nervous system which such injury entails.

Pregnancy.—The effect of pregnancy and the puerperium upon the course of otosclerosis is too well known to require emphasis. In fact, in the writer's experience, its effect has been rather overestimated; and it is not very uncommon to find cases of otosclerosis in which childbearing appears to have had no very detrimental effect on the hearing or on the tinnitus. Still, the fact remains that child-bearing must be placed among the conditions inimical to those who suffer from the disease.

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[1] See chapter on Heredity.

Age.—The effect of increasing years upon otosclerosis is curious and very interesting. The writer has already drawn attention to a fact noticed by him in respect to this matter. In all, at least in almost all cases, no matter whether the deafness has previously progressed rapidly or not, or has remained stationary, there comes a time, usually between the ages of fifty and sixty, when the hearing power begins to diminish rapidly. It is very important to observe that if the hearing tests are applied, it will be found that the increasing deafness is due to changes in the sound-perceiving apparatus rather than in the sound-conducting structures. The explanation of this fact may, perhaps, sometimes be found in the occurrence of arterio-sclerosis, which so frequently manifests itself at this period of life. This explanation, however, fails in many cases, for I have found the rapid increase of the deafness to occur in individuals who had no sign of arterio-sclerosis; and, moreover, it occurs far too uniformly to be capable of such an explanation. A more probable cause of the increasing deafness is to be found in the changes which take place in the nervous system. During youth and early middle life the metabolic changes in the neurone are on the whole anabolic, whereas after that period they become katabolic. A certain amount of internal repair, if the term may be allowed, is possible in the neurones associated with the sense of hearing, during the first half of life, and this may compensate to a certain extent for the destructive changes going on in the neurones in otosclerosis. After this period, however, such compensation disappears, and the hearing power diminishes more rapidly than before.

At first sight it might be thought that this explanation is inconsistent with the fact that in the very young, under 14, the deafness of otosclerosis also proceeds very rapidly, but in these cases the rapid progress of the symptoms is due rather to the changes occurring in the bony capsule than in the sound-perceiving apparatus.

GENERAL TREATMENT

If the reader has carefully considered the section on pathogenesis and etiology, it will be clear, as previously emphasised, that there is no routine treatment for otosclerosis. It is for want of recognition of this fact that the prognosis and treatment of the disease have in the past been so unsatisfactory. Every case must in itself be a special study for the physician. He must spare no pains to ascertain, and estimate accurately, the extent to which the hereditary element is present, or if there is no evidence of such. Any departure from the normal physiological activities of the body must be detected, especially in the alimentary, circulatory, and nervous systems. In addition to these, local conditions in the nose, naso-pharynx, and Eustachian tubes must be studied Finally, it is of the utmost importance to search for any septic focus from which toxins may be absorbed.

It is only after the physician has ascertained the facts in relation to these circumstances that he will be in a position to give an opinion as to whether treatment will be of any avail at all in a given case, and if so, what direction the treatment should take. It may even be necessary to make several examinations before deciding upon a line of action.

It cannot be too strongly emphasised that, before framing a diagnosis and line of treatment, the aurist should ascertain without the possibility of error what is the effect of inflation through the catheter. However clearly satisfied he may be on other grounds that the case is one in which inflation will not produce beneficial results, he must still remember that neither he nor any other physician is infallible, and that it is within the bounds of possibility that the use of the catheter may prove his own diagnosis or that of others to be incorrect.

Assuming that the diagnosis has been correctly made, the physician should, as previously stated, proceed to discover what general or local condition, if any such can be found, may, with a fair degree of probability, be recognised as the exciting cause of the disease. He must then consider whether it is possible to remove this cause or not, and direct his advice and treatment accordingly.

Perhaps the best means of bringing home the value of constitutional treatment and also of demonstrating the truth of the fact that different cases require different constitutional treatment is by the record of cases. I will, therefore, give the following few abbreviated reports from my case-book to illustrate these points.

Cases in which Anæmia was associated with Otosclerosis

Mrs. C., æt. 30, resident in Burmah, consulted me on July 7th, 1912. She noticed three years ago that her hearing was becoming affected. A few months later she began to suffer from buzzing in the ears, which, though constantly present, was of moderate severity. She was somewhat run down in health, but this was not ascribed to anæmia.

She has nine brothers and sisters, and of these, two sisters as well as herself are deaf. Both these sisters have been under my care, and both are subjects of otosclerosis. The father also suffers from the affection.

On examination, it was found that the membranes were normal on both sides.

Rinne: – 7, right; – 9, left.
Watch: 5 ins., right; 4 ins., left.
Whisper: 2½ ft., right; 2 ft., left.

On careful examination in regard to her general condition, it was found that symptoms of anæmia, such as breathlessness, etc., were present, but not in a very marked degree.

Inflation with the catheter produced a slight but very temporary improvement.

The patient was put upon citrate of iron and ammonia internally, and locally a succession of small blisters were applied over the mastoid surface. At the end of two and a half months the patient's health was much improved. The hearing power had risen considerably, as the following figures show:

Watch: right, 1 ft.; left, 9 ins.
Whisper: right, 5 yds.; left, 4½ yds.
Rinne: right, + 2; left, ± 0.

The improvement in this case is very striking, and unfortunately it is rare that such good results can be obtained, especially where there is apparently a certain degree of inherited tendency.

It is interesting also to note that the symptoms of anæmia were not really very marked. This state of matters is, however, not uncommon. That is to say, where there is an individual tendency to otosclerosis, the constitutional change required to transform a potentiality into an actuality need not be very great.

Another case in which anæmia was associated with and presumably causative of otosclerosis is the following:

Miss S. M., æt. 32, consulted me in the year 1902, having become gradually dull of hearing since the age of 26. The family consisted of eight members, and of these there were three deaf, all of whom were examined by myself and found to be subjects of otosclerosis.

The patient had been getting deaf rather more rapidly for a few months before consulting me. Tinnitus was not a noticeable feature in the case. Membranes were not indrawn, but there was a faint rosy tint in both.

Watch: right, 1 in.; left, 1 in.
Whisper: right, 1 ft. +; left, 1 ft.
Conv. voice, right, 4½ yds.; left, 3½ yds.
Rinne: right, − 10; left, − 12.

There was considerable loss of hearing for the low notes in both ears, and for the high notes to a less extent, and also in both ears.

There was a tendency to constipation and anæmia was present, characterised by some degree of breathlessness on exertion; the anæmia, however, was not very pronounced.

Iron was administered for a period of four months, and at the end of that time it was found that the hearing power had risen, the watch being heard at a distance of 1½ ins., and the whisper at 2½ ft. from the right and 1½ ft from the left ear.

It is interesting to note that I had the opportunity of examining this case ten years later, and found that no loss in the hearing had occurred in the interval; and, further, that the ceruminous glands were active, which they had not been at the first examination.

The improvement in this case was distinct, though not so marked as in the former case and in others which I have observed. It is cited rather to show how very lasting the improvement may be, if the treatment is correct.

It must not be supposed that in all cases of otosclerosis associated with anæmia treatment will be as satisfactory as in case 1, or even as in case 2. The following is a case in which the ear affection was associated with anæmia, but which went downhill in spite of the cure of the anæmia.

Mrs. H., æt. 30, complained of dulness of hearing in both ears and noises in the head. She had suffered from the condition for more than six years. The family consisted of eleven members, of whom three were dull of hearing, and on examination all of these proved to be subjects of otosclerosis. The father was dull of hearing at middle life, and on the mother's side two aunts were deaf and had noises in their ears, and in the families of each of these aunts there was at least one member who suffered from deafness and tinnitus.

The patient was anæmic to a marked degree, the breathlessness on exertion being very pronounced, and there was constant tendency to constipation.

There was no indrawing of either membrane, but the faint rosy tint characteristic of some cases of otosclerosis was present. The patient also suffered from paracusis.

Rinne: right, – 12; left, – 9.
Whisper: right, 3 ins.; left, 5 ins.
Watch: only on contact in each ear.

The low notes were markedly lost on both ears.

The administration of iron greatly improved the general condition, and at the end of four months the symptoms of anæmia had disappeared. The noise was still present, and did not appear to have been diminished to any appreciable extent, and the hearing power was, if anything, rather worse. The progress of the deafness may have been lessened in rapidity, but it certainly was not stopped.

This was a case, then, in which the cure of the anæmia did not effect any improvement in regard to the otosclerosis. There are several possible explanations of this result. First, there may have been some other undiscovered constitutional condition which was the exciting cause of the otosclerosis, and which continued to act after the anæmia was cured. Second, the inherent potentiality towards otosclerosis, as evinced by the family tree, may have been so comparatively strong, that when once excited into activity by the anæmia, this activity continued in spite of the cure of the latter. Third, the otosclerosis and the anæmia may have been merely coincident and not correlated in any way. The last is very unlikely, and the second is probably the correct solution.

Cases in which Toxin Absorption from the Alimentary Canal was associated with Otosclerosis

Of all the general conditions which I have found associated with otosclerosis, absorption of toxic substances from the bowel is the most common. It is, however, difficult to say to what extent in a given case the two conditions stand in the relationship of cause and effect. The results of treatment indicate in many cases that the relationship is close, in others it may or may not be so.

Mrs. B., æt. 30, was seen by me on account of dulness of hearing in both ears, of eight years' duration. She suffered from continuous noises in the head, and paracusis was present. She was also the subject of frequent headaches, and of a constant tendency to constipation.

Watch: 2 ins., right; 3½ ins., left.
Whisper: 1 ft., right; 2 ft., left.
Rinne: – 12, right; – 10, left.

The low notes were lost below la$_1$ in both ears. The high notes were heard perfectly. Both membranes were normal, and no rosy tint was present. Inflation produced no improvement either in the hearing or in the tinnitus. Pneumo-massage caused diminution in the tinnitus for a few moments only, and the hearing power was not increased.

Parolein was prescribed in doses of 1 drachm, three times daily, and in the course of a week this was increased to 2 drachms, three times daily. This treatment was continued for four months, and at the end of that time it was found that seven pounds had been gained in weight, the headaches had almost entirely ceased, and there was no longer any necessity for the use of any laxative exept the parolein itself. The hearing power was now as follows:

Watch: 3 ins., right; 5½ ins., left.
Whisper: 2 ft., right; 5 ft., left.

The patient was advised to continue taking the parolein, and at the present time the hearing power is quite as good as at the last note, and the noises are so insignificant that they cause little or no inconvenience. The headaches no longer trouble her.

It is of interest to note that this patient is a sister of Mrs. C., in whom the exciting cause of the disease was anæmia, the cure of which was also followed by considerable improvement. This illustrates in a striking manner the point in regard to the etiology which I have already emphasised, that the exciting cause of otosclerosis is not a specific one. The only single factor which is constant is the innate tendency of the individual.

The next case is reported in order to give an example in which rather marked constipation was present, along with the otosclerosis, but in which the relationship between the two was somewhat doubtful. Further, a third factor, the rheumatic tendency, was present. At any rate, the relief of the constipation was not followed by improvement.

J. McF., æt. 30, a teacher by profession, complained of dulness of hearing in both ears and tinnitus of about nine years' duration. There was a clear family tendency to deafness. There were originally two members of the family of the present generation, but of these the patient alone reached adult life. His mother's family consisted of eight members, all of whom reached adult life; of these, three began to suffer from dulness of hearing, tinnitus, and paracusis between the ages of twenty-five and forty. Of the three, the patient's mother was one. The maternal grandmother also began to suffer from the same symptoms at the age of thirty-nine.

The patient's mother is rheumatic and suffers from constipation. The patient himself has not yet manifested symptoms of rheumatism, but there is a marked tendency to constipation, and, so far as the patient remembers, there always has been. The tinnitus was noticeably worse if the constipation was not relieved.

Watch: not heard at all, right; not heard at all, left.
Whisper: 2–3 ins., right; 9 ins., left.
Rinne: − 8, right; − 10, left.

The hearing power for the high notes was only slightly lost in the right, and not appreciably in the left. The low notes were lost below si$_1$ in both ears. Both membranes were normal in position, but the characteristic rosy tint was present in both. The Eustachian tubes were free from any obstruction, but inflation through the catheter produced no improvement in hearing or in the tinnitus.

The patient was put on paraffin treatment for six months which relieved the constipation and improved the general condition considerably, but it had no effect on the hearing.

In this case there are two possible explanations of the failure of the treatment. The first is that, although the toxic absorption from the bowels was perhaps the exciting cause of the otosclerosis, yet the innate tendency towards the latter was so strong that the pathological changes in the organ of hearing when once roused to activity continued in spite of the removal, to at least a great extent, of the exciting cause. The other is that the constipation was only coincidently present and stood in no causative relationship to the otosclerosis.

Rheumatic manifestations were markedly present in the mother, and it may be that the tendency was present, though latent, in the son, and that the otosclerosis was the first manifestation of its presence. It should be added that anti-rheumatic treatment had been previously given in this case, both by myself and by others, but without producing any improvement. He had also had a course of treatment by Röntgen rays, applied to the ear and adjacent regions, but no benefit accrued.

The following case illustrates again the comparatively beneficial results which may be obtained in these cases of otosclerosis associated with toxic absorption from the alimentary canal, especially if the case is seen early.

Mr. G., æt. 39. Complained of deafness and noises in the ears, the right for ten months and the left for nine years. Of a family of seven, the patient and one sister are deaf, and the sister's symptoms are similar to the patient's. Both parents are alive, and hear well, and there is no known deafness among the aunts, uncles, or cousins, who are fairly numerous. There is a tendency, though not very marked, towards constipation, and the patient occasionally has bilious headaches.

Watch: right, only on contact; left, not heard at all.
Whisper: right, 1 ft.; left, not heard at all.
Conv. voice: right, 6 yds.; left, 1 yd.
Rinne: right, − 5; left, − 10.
Low notes: right, several lost; left, many lost.
High notes: right, all heard; left, lost to a considerable extent.

The right membrane was normal, but the left showed the rosy tint characteristic of otosclerosis. Inflation produced no effect on the left ear, and on the right ear only a very slight improvement which disappeared in a few minutes. Pneumo-massage diminished the tinnitus, but for a period of a few minutes only. A course of blisters over the mastoid for several weeks had no beneficial effect.

A course of liquid paraffin in doses of one tablespoonful at bedtime was then ordered. This treatment was continued for nearly four months, and at the end of that time it was found that some improvement had occurred in the right ear, but none in the left, the measurements then being:

Watch: right, 1 in.; left, not heard at all.
Whisper: right, 2½ ft.; left, not heard at all.

The tinnitus had almost disappeared from the right ear, but in the left it was still distressing. The constipation no longer troubled the patient, he had had no more bilious attacks, and was feeling better altogether.

This illustrates the value of treatment if the case be seen reasonably early. Thus, in the right ear the symptoms had only been apparent for ten months, whereas the left had been affected for nine years. Considerable improvement occurred in the right ear, but none in the left.

The above cases are only a few out of many in which there was reason to suppose that the exciting factor in the production of otosclerosis was toxic absorption from the alimentary tract, and which were treated accordingly, and in many cases with benefit. Before leaving the subject, however, since it is new, I should like to make some further remarks of a more systematic nature.

The idea that toxin absorption from the alimentary canal was frequently related causatively to otosclerosis occurred to me five or six years ago. The close cross-examination of patients as to their general condition *at the time at which the symptoms of otosclerosis first made their appearance, not the time at which the patients were first seen by me*, revealed the fact that in a certain proportion, constipation or other disturbance in the alimentary canal was present. Having gathered a sufficient number of facts to give at least some support to this hypothesis, I proceeded to make an attempt to help these patients by treatment.

The first method was by dieting, and this consisted greatly in diminishing, or even entirely abolishing meat in the diet. A few patients did undoubtedly improve, but the number was small, owing no doubt in part to the fact that many of those treated were unable to give the treatment a fair trial. In some this was due to a lack of self-restraint, in others to domestic difficulties in providing a suitable vegetarian dietary in the household, and in others it was due to the harmful or disagreeable effects which the diet had upon the patients themselves.

The next attempt in the way of treatment was by so-called intestinal antiseptics (salol and β naphthol). These did not prove satisfactory, and were given up.

I then began the administration of liquid paraffin. The method of administration of this substance is so well known that it needs practically no description. I have found it better not to give the drug within an hour of a meal, either before or after. The most suitable method is to give it in one dose, usually a tablespoonful at bedtime, and if more is required, a dessertspoonful or even another tablespoonful may be given in the middle of the morning or of the afternoon. The dosage varies greatly with individuals, and should be ascertained in each case by trial. The test is the effect upon the bowels, for it should render the motions soft but not fluid. In some patients the dose may be less than a tablespoonful daily, while in others it must be considerably augmented.

Under no circumstance have I seen parolein do any real harm to a patient. Its worst effect is the production of flatulence, and such cases are uncommon, and the symptom disappears when the drug is stopped. In some of these individuals, however, the flatulence is so disagreeable that there is no alternative but to give it up. Other methods must then be resorted to, but I know of none nearly so satisfactory in dealing with the disordered condition of the alimentary canal. If parolein suits the patient, it should be administered for a very long time. The treatment should not be given up for many months, at least, and may be continued indefinitely. Some of my patients have taken it for more than a year.

Cases in which Otosclerosis was associated with Chronic Septic Infection

Several cases of otosclerosis have been obser[illegible] me in which the individual potentiality towa[illegible] sclerosis appeared to be rendered active by the ab[illegible] of the toxins of pyogenic organisms.

R. MacC., æt. 39 complained of dulnes[illegible] both ears and noises in the head of a little more [illegible] years' duration. On careful investigation no evi hereditary tendency to deafness was found in the family tree of either the paternal or the maternal side. The patient had suffered from very distressing furunculosis for three years before the onset of the ear affection, and was still suffering when he consulted me. During all this time he stated that he has never been free from at least one furuncle, and frequently several were present coincidently.

Watch: right, 9 ins.; left, 10 ins.
Whisper: right, 3 yds.; left, 3½ yds.
Rinne: right, — 10; left, — 7.
Low notes: lost below ut_1 in both ears.
High notes: heard well, but a few at the top of the scale were lost.

Both membranes were normal in appearance [illegible] rosy tint was present. Inflation showed [illegible] the Eustachian tubes were patent. There was [illegible] improvement after inflation, but this [illegible] course of a few minutes. The treatment adopted was directed towards improving the general health; and locally, a succession of blisters was applied over the mastoid process. Smoking was given up, alcohol was forbidden, and meat in the diet was greatly diminished.

At the end of four months the patient [illegible] himself, but there was no improvement so far as the hearing was concerned. The dulness was not perceptibly worse, but was no better, and the tinnitus was the same as before.

Shortly afterwards the patient, on the advice of his family medical adviser, submitted himself to a course of vaccine treatment for the furunculosis. The case proved to be one of the fortunate ones in which vaccine treatment proved very satisfactory. In the course of a few months he was quite free from furunculosis, and he has remained so since. As regards the ears, the hearing has become no worse during the last two years, and although the tests show no actual improvement, the patient thinks the hearing is a little more acute than it used to be. The tinnitus gradually diminished, and within six or eight months after the cure of the furunculosis it was so insignificant as to give him no trouble at all, whereas formerly it had been intensely irritating.

Miss H., æt. 41, complained of deafness in the left ear of sixteen years' duration and tinnitus of more than two years', and of deafness in the right ear of three years' duration. The left ear, according to the history of the case, became affected as the result of acute middle ear inflammation. The right began to be affected three years ago, a few months subsequent to an attack of appendicitis. The deafness gradually became worse and the patient was treated by several aurists without obtaining any benefit. During this time she had several slight transient attacks of pain in the right iliac region, which were diagnosed as due to appendicitis. A year later she suffered from another attack of appendicitis which was more severe than the first, and six months afterwards a third attack supervened which was so serious as to necessitate immediate operation. She made an excellent recovery, and has remained free from abdominal trouble since. About three or four months after the operation the patient noticed that the tinnitus has disappeared and that the deafness was no longer progressing, rather if anything improving. Neither she nor her medical attendant associated the disappearance of the tinnitus and cessation of the progress of the deafness with the cure of the abdominal trouble.

The case was undoubtedly one of otosclerosis, as (apart even from the family history) the following table shows :

Watch : right, not heard even on contact ; left, not heard even on contact.

Whisper : right, 2 yds. ; left, 6 ins.
Conv. voice : right, 5 yds. ; left, 1 yd.
Rinne : right, − 10 ; left, − 15.
Schwabach : right, + 8 ; left, + 9.
High notes : right, normal ; left, a fair number of notes lost.
Low notes : right, lost below fa$_1$; left, lost below si$_1$.
Weber : to left.

Both membranes were normal in position, and no atrophy or thickening was present. The rosy tint characteristic of otosclerosis was notably pronounced. No improvement resulted from inflation, and the Eustachian tubes were both freely permeable.

In both these cases, therefore, there are reasonable grounds for the belief that septic absorption was the exciting cause of the otosclerosis. In both, the symptoms first made their appearance shortly after septic absorption became established, and in both the tinnitus disappeared and the deafness ceased to progress when the source of the absorption was removed.

In some cases I have a strong suspicion that toxic absorption from a tubercular focus is occasionally the exciting cause of otosclerosis. Two of the cases reported at length in the section on the pathology of otosclerosis were the subjects of phthisis pulmonalis, and the post-mortem examination confirmed the diagnosis. The ear affection in both cases had set in some time after the lungs were diseased, but in neither of these was the tubercular focus removable. Hence, there is no evidence as to the relationship of cause and effect to be derived from the results of. treatment, as in those other cases just reported in which the toxic absorption from the bowel or from septic foci was amenable to treatment.

The question as to whether syphilis, congenital or acquired, is a common exciting cause of otosclerosis is not easy to answer. My own experience is that there is remarkably little evidence in support of a positive answer to this question. To a certain extent the negative results of anti-syphilitic treatment also point in the same direction. Further, the results obtained from the Wassermann test do not indicate any causative relationship between syphilis and otosclerosis. Naturally, syphilis is occasionally present in the subject of otosclerosis, but the proportion does not appear to be perceptibly above that which might be found in a random sample of individuals. It is, however, quite possible that the syphilitic virus may occasionally excite otosclerosis in those in whom the potentiality is markedly present. Different toxins, it has been shown, may do this, and it is difficult to see why the syphilitic poison should be an exception. Clinically, however, as aurists know well, the syphilitic poison, when it affects the hearing, does so in other ways than by the production of otosclerosis. As stated above, I must admit that I have never known anti-syphilitic treatment to improve a case of otosclerosis, and that, too, even in those who were victims of otosclerosis and syphilis coincidently.

From what has gone before the reader will have no difficulty in understanding that in dealing with otosclerosis he is confronted with one of the most difficult and obscure problems in medical science. Probably it is this obscurity which gives to the problem its fascinating interest. It makes a call at once upon the highest intellectual faculties and upon deep human sympathies. The reaction of the disease upon the character and habits of the patient is a study in itself. In some individuals the increasing deafness seems to develop what is best in the character. Shallow and frivolous interests may be gradually replaced by more serious and worthy activities. Unfortunately in some victims the effect of the disease upon the character is not beneficial. Despondency and complaining replace cheerfulness and healthy interests. If the physician is of a sympathetic nature, and is possessed of sufficient personality and gifts, he may have a considerable influence in calling out the better and repressing the less worthy qualities which may make their appearance under the new environment of increasing deafness.

If treatment fails to arrest the progress of the disease, a time comes sooner or later when the patient may require to use an aid to hearing or to learn lip-reading. In respect to both of these, enthusiasts have unwittingly done considerable harm. Some seem to be of opinion that these helps are so satisfactory that they do not take the trouble to make sure that nothing can be done in the way either of improving the hearing or arresting the progress of the disease. More than once I have been consulted by patients who had been recommended to learn lip-reading, when as a result of treatment the hearing improved to an extent sufficient that there was no need for these adventitious methods.

There are, however, not a few cases in which either lip-reading or an aid to hearing is required.

The great drawback to the former is the fact that it is only of use for one purpose, the understanding of the human voice, and even then only under the special condition of direct visual observation. It has the one advantage that no bulky instrument is required. The value of lip-reading is considerably exaggerated by its advocates, especially in these days when great improvements are being made in the manufacture of instruments to aid the deaf.

One of the drawbacks to the use of the various forms of electrophones is the presence of adventitious clanging and jarring sounds. But it may be hoped that in the not distant future this defect will be remedied.

Prophylaxis.—Closely associated with the treatment of otosclerosis is the question of prophylaxis. There is no need to say much in regard to this, however, for the means to be taken to prevent the onset of the disease are practically the same as those employed in treatment, and can be inferred from what has been said in respect to the latter. At the same time it would be ridiculous for the ordinary individual to arrange his way of life with the supreme object of preventing a disease which he will very probably never have, and even never could have; for, as has been shown, it is apparently only certain individuals who have an innate potentiality sufficiently pronounced to favour the onset of the disease.

But when we come to deal with individuals in whom the potentiality is probably marked, as judged by the evidence of otosclerosis in other members of the family, or in the parents or ancestors, the matter is different. It now becomes the duty of the physician to warn the other members of the family, should they consult him, concerning the tendency to otosclerosis, and to advise them as to the best way of living. The individuals may or may not follow his advice, but at any rate the physician has discharged his duty towards them.

With regard to exercise, there is no doubt that very prolonged and violent muscular effort constantly repeated is liable to call into activity the potentiality to otosclerosis. Football, for example, however valuable and healthful to the ordinary individual, is inadvisable for those who have an inherited tendency to the disease. Hockey comes under the same category, and both sexes must be warned against it. Long-distance racing and competitive rowing, as practised at the universities, are also likely to do harm. Tennis and cricket are not likely to be attended with much risk, nor is golf.

In choosing a business or profession, the individuals of a family in which there is any tendency to otosclerosis should be careful. In the first place, the calling should be such that, should deafness occur later in life, it would not be fatal to the pursuance of the calling. I have known cases among barristers, medical men, and stockbrokers in which disaster followed because this point was not taken into consideration. In another case the father of a young man put him into the stockbroking profession against my advice, his mother and several of her brothers and sisters being subjects of otosclerosis. He had to give up his profession three years later, owing to commencing deafness.

In the second place, the profession chosen should be such that overwork and worry are as far as possible avoided; the same must be said with regard to callings which entail much exposure to cold.

The question of the marriage of those who suffer from otosclerosis has already been discussed by me.[1] The aurist is not consulted so frequently as might be expected on this question; a little consideration will show why this is so. One reason is that in an individual who is the subject of otosclerosis, the existence of deafness diminishes his or her chances of marriage.

Should the aurist be consulted about the matter, he must judge each case on its individual merits, and I cannot help thinking that the routine advice to forbid marriage to all these patients is undesirable. As a matter of experience, many marriages of this nature prove extremely happy if the parties are of suitable temperaments.

The question of child-bearing is sometimes brought before the aurist; and here also he must take a wide view of the matter. His answer, to a large extent, must depend upon his opinion as to the effect of child-birth upon the mother in the first place; and the extent to which he fears the occurrence of otosclerosis in the child in the second.

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[1] *Diseases of the Ear*, p. 325 (Baillière, Tindall & Cox)

There is, of course, no doubt that child-birth and the puerperium have a deleterious effect upon otosclerosis in many cases, and it is certainly the physician's duty to warn the woman of this fact. I venture to think that his duty ends there. So far as her own hearing is concerned, the patient is the right one to judge whether she fears more an increase in her deafness or a childless home.

Finally, there sometimes falls upon the physician the duty of answering the question: Is the victim of otosclerosis justified in having children? To answer the question satisfactorily is usually difficult, but in rare cases easy. Unfortunately there is no law of inheritance, as yet discovered, which affords much help in the matter. Mendel's law may be true or not, but the inheritance of otosclerosis, so far as my own investigations go, does not seem to fall within its scope. At any rate, whether the anatomical change characteristic of otosclerosis be considered dominant or recessive, the offspring of those who manifest the defect do not present it in any constant numerical ratio. Of course, in the human subject we are not dealing with pure strains, and perhaps this accounts, in part, for the fact that Mendel's law does not come into evidence. There is also the further difficulty that the law does not take account of environment; and it has been shown (see section on Heredity) that this is of considerable importance, even in cases in which there is a marked tendency to the inheritance of otosclerosis.

The most important factor which will act as a guide in giving advice to a sufferer from otosclerosis, as to whether he or she should have children, is the information derived from the genealogical trees. The worst prospect is where husband and wife both come of stocks in which otosclerosis is found. This is true even where either husband or wife is unaffected by deafness; and, what is still more remarkable, when even both are unaffected. In the latter case, it is very improbable that the physician will be consulted; and if he were, his advice not to have children would probably be disregarded. For it would be difficult to persuade either husband or wife, when both were unaffected by deafness, that there was any great risk of the symptoms occurring in the children.

The condition next in importance is when the wife is herself the subject of otosclerosis, and at the same time comes of a family in which there is a marked tendency to the disease. Under such circumstances there is considerable probability that some of the offspring will become deaf, but a still greater probability that most of them will escape.

When the husband is deaf, and comes of a family with a clear inherited tendency to the disease, the conditions are rather more favourable, the chance of otosclerosis appearing among the offspring being slightly less.

Lastly, when otosclerosis is present in either husband or wife, but there is no other evidence of the disease in either family discoverable over three generations, then the probability of the offspring being affected is slight. In these cases I think it is always wise to advise the individuals to accept the risk.

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